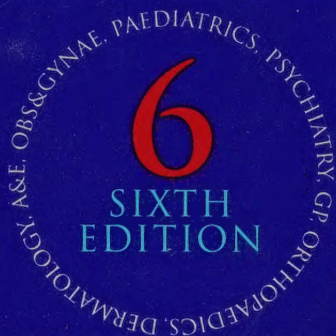


THE DEFINITIVE SPECIALTIES HANDBOOK

OXFORD HANDBOOK OF CLINICAL SPECIALTIES

SIXTH EDITION

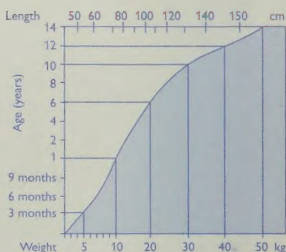


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JUDITH COLLIER
MURRAY LONGMORE
PETER SCALLY

Oral length (cm)	Internal diameter (mm)
18–21	7.5–8.0 (cuffed)
18	7.0 (uncuffed)
17	6.5
16	6.0
15	5.5
14	5.0
13	4.5
12	4.0
	3.5
10	3.0–3.5



Adrenaline (ml of 1 in 10 000) initial intravenous or intraosseous	0.5 1 2 3 4 5
Adrenaline (ml of 1 in 1000) subsequent intravenous or intraosseous (or initial endotracheal)	0.5 1 2 3 4 5
*Atropine (ml of 100 µg/ml) Intravenous or intraosseous (or double if endotracheal)	1 2 4 6 6 6
Atropine (ml of 600 µg/ml)	– 0.3 0.7 1 1 1
Bicarbonate (ml of 8.4%) intravenous or intraosseous (dilute to 4.2% in infants)	5 10 20 30 40 50
*Calcium Chloride (ml of 10%) intravenous or intraosseous	0.5 1 2 3 4 5
Diazepam (ml of 5 mg/ml emulsion) intravenous or rectal	0.4 0.8 1.6 2 2 2
Diazepam (mg rectal tube solution) rectal	2.5 5 10 10 10 10 mg
Glucose (ml of 50%) intravenous or intraosseous (dilute to 25% in infants)	5 10 20 30 40 50
*Lignocaine (ml of 1%) intravenous or intraosseous	0.5 1 2 3 4 5
Naloxone neonatal (ml of 20 µg/ml) intravenous or intraosseous	2.5 5 – – – –
Naloxone adult (ml of 400 µg/ml)	– 0.25 0.5 0.75 1 1.25
*Salbutamol (mg nebuliser solution) by nebuliser (dilute to 2.5–5 ml in physiological saline)	– 2.5 5 5 5 5 mg
Initial DC defibrillation (j) for ventricular fibrillation or pulseless ventricular tachycardia	10 20 40 60 80 100j
Initial DC cardioversion (j) for supraventricular tachycardia with shock (synchronous) or ventricular tachycardia with shock (non synchronous)	5 5 10 15 20 25j
Initial fluid bolus in shock (ml) crystalloid or colloid	100 200 400 600 800 1000

*Caution! Non-standard drug concentration may be available;

Use **atropine** 100 µg/ml or prepare by diluting 1 mg to 10 ml or 600 µg to 6 ml in physiological saline.

Note that 1 ml of **calcium chloride** 10% is equivalent to 3 ml of **calcium gluconate** 10%.


Use **lignocaine** (without adrenaline) 1% or give twice the volume of 0.5%; give half the volume or 2% or dilute appropriately.

Salbutamol may also be given by slow intravenous injection (5 µg/kg), but beware the different concentrations available (eg. 50 and 500 µg/ml).

†P Oakey 1993 *BMJ* 306 1613 Reproduced by kind permission

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Oxford Handbook of Clinical Specialties

SIXTH EDITION

JUDITH COLLIER
MURRAY LONGMORE
PETER SCALLY

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Preface to the sixth edition

There have been three great changes in the way we have viewed ourselves: the first happened when we realised that we were not at the centre of the universe; the second happened when we realised, with Darwin, that we are here by accident, thanks to natural selection, the survival of the fittest, and the 'wasteful, blundering low & horridly cruel works of nature'. The latest change relates to that little word 'we' in the sentences above. We are changing *ourselves*. When John Donne complained in 1611 that the newly designed firmament was casting everything into question

And new Philosophy calls all in doubt,
The element of fire* is quite put out;
The sun is lost, and th'earth, and no man's wit
Can well direct him, where to look for it.
And freely men confess, that this world's spent,
When in the Planets, and the Firmament
They seek so many new; they see that this
Is crumbled out again to his Atomis.
'Tis all in pieces, all coherence gone;
All just supply, and all relation:
Prince, subject, father, son, are things forgot

*i.e. the sun

He was exaggerating: he took his life and death for granted, he could not add stature to his offspring (no genetically engineered growth hormone, p 759); he was secure in the dichotomy of all humans into male and female (he had no instrument to measure shades of gender, p 222), and he was secure in the knowledge that the coming together of male and female was necessary to the begetting of life. Now we have human cloning (p 57), gene therapy (p 201), the transplanting of ovaries (p 57) and even children with two genetic mothers—one for cytoplasmic DNA and the other for nuclear DNA (see *ooplasmic transfer*, p 57). When cells deliberately infected with apparently harmless retroviruses carrying special genes are implanted into our marrow, new diseases can be treated and old questions about identity become irrelevant (see the therapy of severe combined immunodeficiency, p 201). If we have a few mouse cells secreting dopamine in my substantia nigra our thoughts are still our own: but how many mice does it take before we get dizzy looking into the heart of this question? All these developments are faithfully reproduced in these pages—often in parentheses, in footnotes, and in obscure places. For the truth is that all these interesting developments are *at present*, irrelevant to health. The infectious diseases which kill most children are only partly the result of microbes; they are equally the product of human failings, and these show no signs of changing. Water-borne diseases spread not because of certain biological facts, but because of man-made events, and, above all, by age-old human emotions: greed for money, the race to exploit our world, and the desire to dominate (war makes refugees, poverty, and dislocation—the ideal breeding ground for new diseases). These are also not mentioned much in these pages (but see p 187): this is not because they have been hived off to parentheses, footnotes and so on: it is rather that these emotions are the ink (or blood) with which every word of this volume is written.

Judith Collier & Murray Longmore—*Ferrin*
Peter Scally—*Mater Hospital, Queensland and University of Queensland*

Drugs

While every effort has been made to check this text, it is still possible that drug or other errors have been missed. Also, dosage schedules are continually being revised and new side-effects recognized. Oxford University Press makes no representation, express or implied, that drug dosages in this book are correct. For these reasons, the reader is strongly urged to check with the most up to date published product information and *Data sheets* provided by the manufacturers, and the most recent codes of conduct and safety regulations. The authors and the publishers do not accept responsibility or legal liability for any errors in the text, or for the misuse or misapplication of material in this work.

Except where otherwise stated, drug doses and recommendations are for the **non-pregnant adult** who is **not breast feeding**.

Readers are reminded of the need to keep up to date, and that this need can only ever be *partly* addressed by printed texts such as this.

Drug nomenclature

This book uses British^{uk} approved names, followed, where there is a difference, by the recommended international non-proprietary name (rINN; usually there is no difference) as stipulated by European Directive. Exceptions to our rule of giving both names occur where the change is minute, eg amoxycillin^{uk}/amoxicillin, where the rINN is used, to avoid tedious near-duplications. Among the new rINNs used are:

Alimemazine	(trimeprazine ^{uk})	Epinephrine	(adrenaline ^{uk})
Amoxicillin	(amoxycillin ^{uk})	Furosemide	(frusemide ^{uk})
Bendroflumethiazide	(bendrofluazide ^{uk})	Lidocaine	(lignocaine ^{uk})
Ciclosporin	(cyclosporin ^{uk})	Chlormethine	(mustine ^{uk})
Clomifene	(clomiphene ^{uk})	Norepinephrine	(noradrenaline ^{uk})
Diethylstilbestrol	(stilboestrol ^{uk})	Sulfonamides (all)	(sulphonamides ^{uk})
Dosulepin	(dothiepin ^{uk})	Trihexyphenidyl	(benzhexol ^{uk})

In addition, rINNs for cephalosporins (or cephalosporins) now all start with *cef.* . . (not *ceph*). For consistency, and to avoid distracting variations in adjacent usages, we have spelt sulphur (and its derivatives) *sulfur* as all *sulphonamides* have a rINN starting *sulf.* . . Problems remain with oestrogens such as ethinyl(o)estradiol and (o)estradiol: here we use the shorter spelling, as far as drugs are concerned, so that we abide by the directive, while retaining the uk spelling of oestrogens in non-pharmacological contexts.

Preface to the first edition

When someone says that he is 'doing obstetrics'—or whatever, this should not hide the fact that much more is being done besides, not just a little of each of medicine, psychiatry, gynaecology and paediatrics, but also a good deal of work to elicit and act upon the patient's unspoken hopes and fears. At the operating table he must concentrate minutely on the problem in hand; but later he must operate on other planes too, in social and psychological dimensions so as to understand how the patient came to need to be on the operating table, and how this might have been prevented. All the best specialists practise a holistic art, and our aim is to show how specialism and holism may be successfully interwoven, if not into a fully watertight garment, then at least into one which keeps out much of the criticism rained upon us by the proponents of alternative medicine.

We hope that by compiling this little volume we may make the arduous task of learning medicine a little less exhausting, so allowing more energy to be spent at the bedside, and on the wards. For a medical student coming fresh to a specialty the great tomes which mark the road to knowledge can numb the mind after a while, and what started out fresh is in danger of becoming exhausted by its own too much. It is not that we are against the great tomes themselves—we are simply against reading them too much and too soon. One starts off strong on 'care' and weak on knowledge, and the danger is that this state of affairs becomes reversed. It is easier to learn from books than from patients, yet what our patients teach us may be of more abiding significance: the value of sympathy, the uses of compassion and the limits of our human world. It is at the bedside that we learn how to be of practical help to people who are numbed by the mysterious disasters of womb or tomb, for which they are totally unprepared. If this small book enables those starting to explore the major specialties to learn all they can from their patients, it will have served its purpose—and can then be discarded.

Because of the page-a-subject format, the balance of topics in the following pages may at first strike the reader as being odd in places. However, it has been our intention to provide a maximally useful text, rather than one which is perfectly balanced in apportioning space according to how common a particular topic is—just as the great *Terrestrial Globes* made by George Phillips in the 1960s may seem at first to provide an odd balance of place names, with Alice Springs appearing more prominently than Amsterdam. To chart a whole continent, and omit to name a single central location out of respect for 'balance' is to miss a good opportunity to be useful. George Phillips did not miss this opportunity, and neither we hope, have we. It is inevitable that some readers will be disappointed that we have left out their favoured subjects (the Phillips' Globe does not even mention Oxford!). To these readers we offer over 300 blank pages by way of apology.

Ferring
1987

J.A.B.C.
J.M.L.

Conflicts of interest: none declared

Because of numerous and well-publicized occasions where writers of guidelines recommending certain drugs turn out to have undisclosed financial contacts with the pharmaceutical industries concerned, we wish to place on record that we have no contacts with any pharmaceutical company, and no pharmaceutical company employs us in any capacity, and neither have we received any financial input bearing upon our research for this publication. We have a policy of not seeing representatives from the pharmaceutical industry, or receiving their gifts or hospitality. We assert that the drugs recommended in this book have been selected on the basis of the best available evidence.

J.A.B.C. & J.M.L., principal authors

Symbols and abbreviations

	This is important	CT	Computer tomography
	Don't dawdle! Prompt action saves lives	CVP	Central venous pressure
	Reference to evidence-based medicine (see p 433).	CVS	Cardiovascular system
	More details in the electronic version of this text (p 446)	CXR	Chest x-ray
	Conflict (controversial topic)	D	Dimension
$\Delta\Delta$	Differential diagnosis	D&C	Dilatation (cervix) & curettage
σ/φ	Male to female ratio	dB	Decibel
#	Fracture	DIC	Disseminated intravascular coagulation
\therefore	On account of	DIP	Distal interphalangeal
$\sim ; =$	About; approximately equal to	dL	Decilitre
[@1234...]	Search medline (p 447) with the number after '@' to find the abstract referred to	DM	Diabetes mellitus
-ve/+ve	Negative/positive	DoH	Department of Health
$\downarrow ; \uparrow$	Decreased; Increased	D&V	Diarrhoea and vomiting
\leftrightarrow	Normal (eg plasma level)	DVT	Deep venous thrombosis
A&E	Emergency department	E-BM	<i>Evidence-based Medicine</i> (journal)
ac	Ante cibum (before food)	EBV	Epstein-Barr virus
ACE(i)	Angiotensin-converting enzyme (inhibitor)	ECG	Electrocardiogram
ACLS	Advanced cardiac life support	ECT	Electroconvulsive therapy
ACTH	Adrenocorticotrophic hormone	EEG	Electroencephalogram
ADH	Antidiuretic hormone	ENT	Ear, nose and throat
AFP	α -fetoprotein	ESR	Erythrocyte sedimentation rate
AIDS	Acquired immuno-deficiency syndrome	ET	Endotracheal
Alk	Alkaline (phos = phosphatase)	FB	Foreign body
ALT	Alanine aminotransferase	FBC	Full blood count
ANF	Antinuclear factor	FH	Family history
AP	Anteroposterior	FSH	Follicle-stimulating hormone
APH	Antepartum haemorrhage	G	Gauge
A(P)LS	Advanced (Paediatric) Life Support <i>Manuals</i> (269)	g	Gram
ASO	Antistreptolysin O (titre)	G6PD	Glucose-6-phosphate dehydrogenase
ASD	Atrioseptal defect	GA	General anaesthesia
ATLS	Advanced Trauma Life Support <i>Manual</i> reference	GI	Gastrointestinal
AV	Atrioventricular	GP	General practitioner
AVM	Arteriovenous malformation	G(γ)GT	Gamma (γ) glutamyl transpeptidase
BMJ	<i>British Medical Journal</i>	GU	Genitourinary
BP	Blood pressure	h	Hour
BJGP	<i>British Journal of General Practice</i>	Hb	Haemoglobin
BNF	<i>British National Formulary</i>	HBsAg	Hepatitis B surface antigen
ca	Carcinoma	HDL	High-density lipoprotein
CCF	Combined (right & left sided) cardiac failure	HIV	Human immunodeficiency virus
ChVS	Chorionic villus sampling	HRT	Hormone replacement therapy
CI	Contraindications	HVS	High vaginal swab
CMV	Cytomegalovirus	Ib/ibid	Ibidem (in the same place)
CNS	Central nervous system	ICP	Intracranial pressure
CoC	Combined oral contraceptive	IE	Infective endocarditis
CPR	Cardiopulmonary resuscitation	Ig	Immunoglobulin
CRP	C-reactive protein	IHD	Ischaemic heart disease
CSF	Cerebrospinal fluid	IM	Intramuscular
		INR	International normalized ratio of prothrombin time
		IQ	Intelligence quotient
		IP	Interphalangeal
		IPPV	Intermittent positive pressure ventilation
		ITP	Idiopathic thrombocytopenic purpura

ITU	Intensive therapy unit	PDA	Patent ductus arteriosus
iu	International unit	PET	Pre-eclamptic toxæmia
IUCD	Intrauterine contraceptive device	PID	Pelvic inflammatory disease
IV	Intravenous	PIP	Proximal interphalangeal
IVI	Intravenous infusion	PO	Per os (by mouth)
IVU	Intravenous urography	PoP	Progesterone-only pill
JVP	Jugular venous pressure	PPH	Postpartum haemorrhage
K⁺	Potassium	PR	Per rectum
kg	Kilogram	PTR	Prothrombin ratio
kPa	Kilopascal	PE	Pulmonary embolism
L	Litre	PUO	Pyrexia of unknown origin
LA	Local anaesthesia	PV	Per vaginam (latin)
LDH	Lactate dehydrogenase	RBC	Red blood cell
LFT	Liver function test	RCGP	Royal College of General Practitioners
LH	Luteinizing hormone	RA	Rheumatoid arthritis
LMP	Day 1 of last menstrual period	RTA	Road traffic accident(s)
LP	Lumbar puncture	RUQ	Right upper quadrant
LVH	Left ventricular hypertrophy	RVH	Right ventricular hypertrophy
µg	Microgram	R_x	Treatment (prescribing drugs)
MAOI	Monoamine oxidase inhibitor	S₁; S₂	1 st and 2 nd heart sounds
MCP	Metacarpophalangeal	SBE	Subacute bacterial endocarditis
MCV	Mean cell volume	SC	Subcutaneous
medline	(See p 447); when followed by ~, the link is a full-text link	SCBU	Special care baby unit
mg	Milligrams	SE	Side-effects
mL	Millilitre	sec	Seconds
mmHg	Millimetres of mercury	SLE	Systemic lupus erythematosus
MRI	Magnetic resonance imaging	SaO₂	Oximetry estimation of capillary O ₂ saturation; also SaO ₂
MSU	Midstream urine culture	SSRI	Selective serotonin reuptake inhibitors
MTP	Metatarsophalangeal	STD	Sexually transmitted disease
N=228*	Medline (p 447) reference to a randomized trial of 228 patients (*or whatever number follows N)	SVC	Superior vena cava
n = 98*	Medline reference to a non-randomized trial of 98 patients	t_½	Half life
NaCl	Sodium chloride	T_°	Temperature
NBM	Nil by mouth	T₃	Triiodothyronine
NEJM	<i>New England Journal of Medicine</i>	T₄	Thyroxine
NGT	Nasogastric tube	TB	Tuberculosis
NHS	National Health Service	TIA	Transient ischaemic attack
NICU	Neonatal intensive care unit	TFT	Thyroid function tests
N₂O	Nitrous oxide	TPR	Temperature, pulse, and respirations
NSAIDs	Non-steroidal anti-inflammatory drugs	TSH	Thyroid-stimulating hormone
OED	<i>Oxford English Dictionary</i> 1 st ed	u (or U)	Units
OHCM	<i>Oxford Handbook of Clinical Medicine</i> 5 th edition,oup	U&E	Urea and electrolytes
ORh-ve	Blood group O, Rh negative	UK	United Kingdom
PA	Posteroanterior	URTI	Upper respiratory tract infection
PAN	Polyarteritis nodosa	US(S)	Ultrasound (scan)
P_aCO₂	Partial pressure of CO ₂ in arterial blood	UTI	Urinary tract infection
P_aO₂	Partial pressure of oxygen in arterial blood	UV	Ultraviolet
pc	Post cibum (after food)	VSD	Ventriculoseptal defect
PCV	Packed cell volume	WCC	White blood cell count
		wt	Weight
		 yrs	Years

Other abbreviations are given in full on the pages where they occur.

Night-visit with Nietzsche

In a small car on an open road, at night, I am heading to a sick child, aged 10. Nietzsche is sitting beside me, it seems, and muttering in the dark: 'Why bother with all this medicine? It gives you no pleasure, and I see you feel no compassion. Be a free spirit like me! Allow me to say to you "You are entangled in an unyielding snare and straightjacket of duties and cannot get free". You think you are making a sacrifice to a noble cause; but listen to this: "Anyone who has truly offered a sacrifice knows that he wanted something for it and got it—perhaps something of himself in return for something of himself—that he gave up something here in order to have more there, perhaps just to be more, or at least to feel as if he were more".'

I drive on—obediently strapped into my straightjacket of duties. Then, on my right hand, I see, between two low houses a huge moon, rising, right between them, low and red. It's far too big to believe—but I know I have seen this sight once before—aged 10—a moon rising over the river Cherwell. Nietzsche cannot see it. He's too high. But I'm slumped over the wheel; I can see it just as it was a few decades ago. I wax. I wane. Ideals come and go, and will return. I do my night visit in a strange calm, and, on my way back, I look for my moon, to show it to Nietzsche. But Nietzsche has gone, and the moon is shining on someone else's night visit. Your night visit perhaps? I will wear your straightjacket and you can wear mine. I've got my moonlight inside me now; enough for a few more decades, I wonder?

When I last looked, we were walking through the moonlight, singing in our chains.

See Friedrich Nietzsche *Beyond Good and Evil* (aphorisms 220–226) and the works of Maura D St John.

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Pronouns For brevity, the pronoun 'he' or 'she' has been used in places where 'he or she' would have been appropriate. Such circumlocutions do not aid the reader in forming a vivid visual impression, which is one of the leading aims of good authorship. Therefore, for balance and fairness and where sense allows, we have tried alternating *he* with *she*.

Understanding our patients

Most of the time we treat our patients quite well, without ever really understanding them. The idea that we should strive to understand and empathize with *all* our patients is ultimately unsustainable. Out-patient clinics and surgeries would grind to a halt, and urgent visits would never get done. It is also possible that to do so would be counter-productive from the patient's point of view. For two human beings to understand each other's inner life is a rare event, and if we offered this understanding to all our patients they might become addicted to us, and be unable to get on with the rest of their lives. Nevertheless, it is good practice to try to understand *some* patients. Doing so may entail swallowing an alien world and digesting it rather slowly. Paradoxically, to achieve this, we very often need to keep our mouths *shut*, particularly with those in whom we have reached a therapeutic impasse—for example if the illness is untreatable, or the patient has rejected our treatment, or if the patient seems to be asking or appealing for something more. Eye contact is important here. One of the authors (JML) recalls forever his very first patient—found on a surgical ward recovering from the repair of a perforated duodenal ulcer: a nice simple surgical patient, ideal for beginners. I asked all the questions in the book, and knew all his answers and his physical features: even the colour of his eyes. Luckily, the house officer who was really looking after him did not ask so many questions, and knew how to interpret the appeal for help behind those eyes, and in his busy day found space to receive the vital clue beyond my grasp—that my patient was a drug addict and under great stress as he could no longer finance his activity.

So, the first step in trying to understand a patient is to sit back and listen. Next, if possible, it is very helpful to see your patient often, to establish rapport, and mutual respect. If the relationship is all one way, with the doctor finding out all about the patient, but revealing nothing of him- or herself, this mutual respect can take a very long time to grow. But beware of sharing too much of your own inner life with your patients: you may overburden them, or put them off. Different patients respond to different approaches. Understanding patients inevitably takes time, and it may be hard in a series of short appointments. A visit to the patient's home may be very revealing, but for many doctors trapped in hospital wards or clinics, this is impossible. But it is usually possible to have a longish, private interview, and take whatever opportunity arises. We once worked with a consultant who infuriated his junior staff on busy ward rounds by repeatedly selecting what seemed to us the most boring and commonplace medical 'cases' (such as someone with a stroke) and proceeding to draw the curtain around the patient's bed to exclude us, and engage in what seemed like a long chat with the patient, all in very hushed voices, so that we never knew what he said—until Sister told us that he never said anything much, and simply received anything that was on the patient's mind. For the most part, he was swallowing their world in silence. We came to realise that there was nothing that these patients, robbed as they were of health and wholeness, appreciated more in their entire hospital stay.

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A holistic approach to gynaecology: Parry's* dictum

It is more important to know what sort of patient has the disease than what kind of disease the patient has.

*Caleb Parry (1755–1822) was a doctor in Bath who was fascinated by the events in his patients' lives, and their connection with their diseases—some of which he described for the first time. These early descriptions are remarkable for their effortless intertwining of psychological and physical phenomena. www.thyroidhistory.net/hyperthyroidism.html

All this suggests a rather serious, one-sided doctor, but this is wrong. To get away from it all, he became a keen balloonist, lending ideas and materials to Edward Jenner (who dedicated his magnum opus on smallpox to him). From the Royal Crescent in Bath, he launched his great balloon, which, like his bedside manner, was made of varnished silk.

His aphorism is particularly relevant to this chapter because many of its diseases are chronic, and the choices of treatment are many. Take endometriosis (p 52), for example. If an examiner were to ask you 'what is the treatment for endometriosis'—you might well look at him thoughtfully, before replying that it all depends on who has got it—where they are in their lives, how much the pain matters, what the plans are for future pregnancies, how these plans may be ambiguous and change, according to work, relationships, and the onset of friendships. What does the patient feel about long-term medication with agents which can change her sexuality, and hence the person who is suffering the disease? Some may tolerate doctor-induced hypoestrogenism (flushes, decreased libido, loss of bone density) thinking the price well worth paying for relief of endometriosis symptoms; others will take the opposite view; in a few, their ability to take a decision will be influenced by the drugs they are already taking. [medline.6682476](https://pubmed.ncbi.nlm.nih.gov/6682476/)

Sometimes rational choice is the hardest thing '...how can you expect me to make a rational choice until you sort out these dreadful periods of mine: I cannot even think...'

In summary: ►take time ►understand your patient ►offer all options...then... ►let her choose.

History and examination


History Let her tell the story, but remember that she may be reluctant to admit some problems, particularly if you are a man, so make sure to cover them in your questions.

- 1 **Menstrual history:** ► Note date of last menstrual period (LMP), ie first day of bleeding, or menopause. Was the last period normal? Cycles: number of days bleeding/number of days from day 1 of one period to day 1 of next (eg 5/26). Are periods regular? If heavy, are there clots or flooding? How many pads/tampons are needed (this may not be a reliable guide)? Are periods painful? Is there bleeding between periods, postcoitally, or since the menopause? Age at menarche?
- 2 **Obstetric history:** How many children? Antenatal problems, delivery, gestation, outcome (and weights of babies) and puerperium of each pregnancy? Terminations and miscarriages—at *what* stage, *why*, and (terminations) *how*?
- 3 **Symptoms:** If she has *pain* what is it like? Uterine pain tends to be colicky and felt in the sacrum and groins. Ovarian pain tends to be felt in the *iliac fossa* and radiates down front of the thigh to the knee. Ask about *dyspareunia* (painful intercourse). Is it superficial (round the outside) or deep inside? If she has *vaginal discharge* what is it like (amount, colour, smell, itch); when does she get it? Ask about *prolapse* and *incontinence*. When? How bad? Worse whilst standing? Ask about bowel symptoms (irritable bowel can cause pelvic pain), faecal incontinence.
- 4 **Sex and contraception:** Is she sexually active? Are there physical or emotional problems with sex? What contraception is she using and is she happy with it? What has she tried previously? Has she had problems conceiving? If so, has she had treatment for infertility? What about sexually transmitted diseases? Date and result of last smear?
- 5 **Other:** General health, smoking. Previous gynaecological treatment.

Examination ► Many women find pelvic examination painful and undignified, and particularly embarrassing if you are male. Explain what you are going to do. Be gentle. Use a chaperone.

General: Is she well or ill? Is she shocked? If so, treat it.

Abdomen: Look for tenderness and peritonism. If there is a mass, could it be a pregnancy? Listen for a fetal heart (p 84).

Vaginal examination: (p 6). Use your eyes to inspect the vulva, a *speculum* to examine the vagina and cervix and your *fingers* to assess the uterus and adnexae bimanually. Examination is usually done with the patient on her back or in the left lateral position (preferable for detecting prolapse). *Sims' speculum* has 2 right-angle bends, and is used for inspecting the vaginal walls, eg for prolapse and incontinence. 

Cusco's (bivalve) speculum is used for inspecting the cervix with the aid of a light. Insert the speculum closed (warmed under a tap) with the blades parallel to the labia; lubricating jelly makes it easier. When it is in, rotate it and open it. The speculum should achieve its full length before opening and usually the cervix will pop into view. If it does not, do a bimanual to check the position of the cervix and try again. Do swabs (p 48) and a cervical smear (p 32) if indicated. Close the speculum gradually, under direct vision, as you withdraw it, to avoid trapping the cervix.

Sexual health

Sexual health is the enjoyment of sexual activity of one's choice, without causing or suffering physical or mental harm.¹ Of course there is more to sex than enjoyment. 'Perhaps the sexual life is the great test. If we can survive it with charity to those we love, and affection to those we have betrayed, we needn't worry so much about the good and the bad in us. But jealousy, distrust, cruelty, revenge, recrimination. . . then we fail. The wrong is in that failure even if we are the victims and not the executioners. Virtue is no excuse. . .'²

Once one understands that human sexuality is infinitely complex, it is easier to appreciate statistics such as 'sexual dysfunction is a big health problem, affecting 43% of women and 31% of men'.³

Enemies of sexual health include:

- Disharmony in personal relationships, or simply *too many* relationships.
- Pain, or any medical, or gynaecological condition.
- Anxiety (whether or not related to fear of failure); depression; fatigue.
- Drugs (eg tamoxifen; the Pill; cyproterone; antidepressants; narcotics).
- A multiplicity of irreconcilable rôles (if your patient is trying to achieve ascendancy in her work, as well as being chief shopper, cook, housewife, mother, and friend, then the rôle of lover may be eclipsed—all the more so if she also finds herself in the rôle of being chief person to blame if things go wrong—if the fridge is empty, if the money runs out, if the children do not get to school on time, or if her partner loses his job).
- Myths about sexual performance (eg that all physical contact must lead to sex, that sex equals intercourse, and that sexual relations should come naturally and easily).⁴

Gynaecological anatomy

The vulva comprises the entrances to the vagina and urethra, the structures which surround them (clitoris, labia minora, and fourchette), and the encircling labia majora and perineum. The hymen, when broken (by tampons, or intercourse) leaves tags at the mouth of the vagina.

Look for: Rashes; atrophy; ulcers; lumps (p 28–30); deficient perineum (you can see the back wall of the vagina); incontinence.

The vagina is a potential space with distensible folded muscular walls. The contents of the rectum, which runs behind the posterior wall, may be palpated through the vagina. The cervix projects into the vault at the top which forms a moat around it, deepest posteriorly, conventionally divided into anterior, posterior, and two lateral fornices. From puberty until the menopause lactobacilli in the vagina keep it acidic, discouraging infection.

Look for: Inflammation; discharge (p 48); prolapse (p 54).

The cervix is mostly connective tissue. It feels firm, and has a dent in the centre (the opening, or os, of the cervical canal). Mucin-secreting glands of the endocervix lubricate the vagina. The os is circular in nulliparous women, but is a slit in the parous.

Look for: Pain on moving the cervix (excitation—p 24 and p 50); ectopy; cervicitis and discharge; polyps, carcinoma (p 32).

The uterus has a thick muscular-walled body lined internally with columnar epithelium (the endometrium) connected to the cervix or neck. It is supported by the uterosacral ligaments. The peritoneum is draped over the uterus. The valley so formed between it and the rectum is the rectovaginal pouch (of Douglas), and the fold of peritoneum in which the Fallopian tubes lie is known as the broad ligament. The size of the uterus is by convention described by comparison with its size at different stages of pregnancy. Since that is variable, estimates are approximate, but the following is a guide: nonpregnant—plum-sized; 6 weeks—egg; 8 weeks—small orange; 10 weeks—large orange; 14 weeks—fills pelvis.

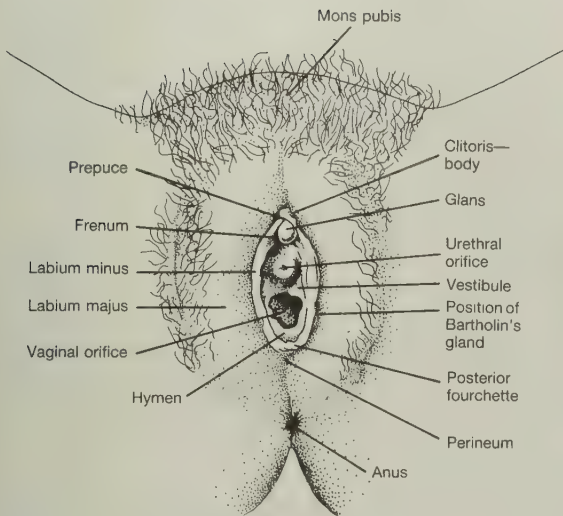
In most women the uterus is *anteverted*, ie its long axis is directed forward and the cervix points backwards. The body then flops forwards on the cervix—*anteflexed*. An anteverted uterus can be palpated between the two hands on bimanual examination (unless the woman is obese or tense) or the bladder is full.

In 20% it is *retroverted and retroflexed* (p 6).

Look for: Position (important to know for practical procedures); mobility (especially if retroverted); size; tenderness (p 24 and p 50).

Adnexae These are the *Fallopian tubes*, *ovaries*, and associated connective tissue (parametria). They are palpated bimanually in the lateral fornices, and if normal cannot be felt. The ovaries are the size of a large grape and may lie in the rectovaginal pouch.

Look for: Masses (p 42) and tenderness (p 50).



Genital abnormalities

Vagina and uterus These are derived from the Müllerian duct system and formed by fusion of the right and left parts. Different degrees of failure to fuse lead to duplication of any or all parts of the system.

Vaginal septae are quite common (and often missed on examination).

Duplication of the cervix and/or uterus may also be missed, eg until the woman becomes pregnant in the uterus without the iucp!

A partially divided (*bicornuate*) uterus or a uterus where one side has failed to develop (*unicornuate*) may present as recurrent miscarriage particularly in the second trimester, or as difficulties in labour. Such abnormalities are diagnosed by *hysterosalpingogram*.

An absent uterus or a rudimentary uterus with absent endometrium is rare. They present with primary amenorrhoea.

An absent or short vagina is uncommon but can be corrected by plastic surgery. The membrane at the mouth of the vagina where the Müllerian and urogenital systems fuse (the hymen) may be imperforate. There is apparent primary amenorrhoea, with a history of monthly abdominal pain and swelling, and the membrane bulging under the pressure of dammed up menstrual blood (*haematocolpos*). It is relieved by incising the membrane. NB: in some communities, female 'circumcision'* is still practised, and this is another cause of *haematocolpos*.

6

►Renal system abnormalities often coexist with genital ones, so ivu and ultrasound should be performed.

Ovary Thin, rudimentary 'streak' ovaries are found in Turner's syndrome (p 759). Ovaries are absent in testicular feminization syndrome, but primitive testes are present (p 222). Remnants of developmental tissue (eg the Wolffian system) may result in cysts around the ovary and in the broad ligament.

Uterine retroversion

About 20% of women normally have a retroverted retroflexed uterus which is fully mobile. It is difficult to palpate bimanually unless you can push it into anteversion by pressure on the cervix. It causes no problems except (rarely) if it fails to lift out of the pelvis at 12 weeks of pregnancy, presenting with discomfort and retention of urine, eg at 14 weeks; catheterization and lying prone may relieve it.

Inflammation in the pelvis (due to infection or endometriosis) can cause adhesions which tether the uterus in a retroverted position. The patient may present with dysmenorrhoea, dyspareunia, or infertility—problems which can only be relieved by treatment of the underlying disease.

*Circumcision is a misnomer, as it implies a parallel with male circumcision, which is incorrect: the female 'operation' is carried out later (at 5–7yrs) and may be much more damaging and extensive, eg as practised in Somalia. Procedures include clitoridectomy ± infundibulation (removal of the clitoris, labia minora and labia majora, with oversewing or aposition by thorns). Even the milder Sunna entails removal of part of the clitoris, and more may be removed than intended. At betrothal, an attendant cuts the scar tissue to allow intercourse. Long-term problems include very slow urination, *haematocolpos*, dyspareunia or non-consummation, obstructed labour, as well as emotional trauma. See H Gordon 1998 *Diplomate* 5 86.

Normal menstruation

Puberty is the development of adult sexual characteristics. The sequence: breast buds develop, then growth of pubic hair, then axillary hair. Then menses begin (**menarche**) from 10½ years onwards (mean ~13yrs; age was falling in the UK, but no longer); it is earlier if short and overweight; investigate if no periods by 16yrs (p 10). A growth spurt (p 200) is the first change in puberty and is usually completed 2yrs after menarche when the epiphyses fuse.

The menstrual cycle The cycle is controlled by the 'hypothalamic-pituitary-ovarian (HPO) axis'. Pulsatile production of gonadotrophin-releasing hormones by the hypothalamus stimulates the pituitary to produce the gonadotrophins: follicle stimulating hormone (FSH) and luteinizing hormone (LH). These stimulate the ovary to produce oestrogen and progesterone. The ovarian hormones modulate the production of gonadotrophins by feeding back on the hypothalamus and pituitary.

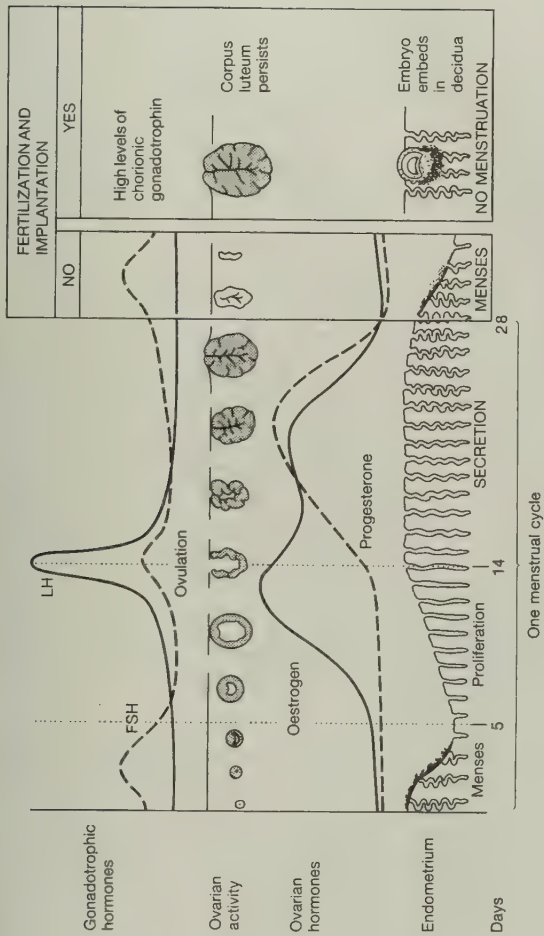
Day 1 of the cycle is the first day of menstruation. Cycle lengths vary greatly; only 12% are 28 days. Cycles soon after menarche and before the menopause are most likely to be irregular and anovulatory. In the first 4 days of the cycle, FSH levels are high, stimulating the development of a primary follicle in the ovary. The follicle produces oestrogen, which stimulates the development of a glandular 'proliferative' endometrium and of cervical mucus which is receptive to sperm. The mucus becomes clear and stringy (like raw egg white) and if allowed to dry on a slide produces 'ferning patterns' due to its high salt content. Oestrogen also controls FSH and LH output by positive and negative feedback.

14 days before the onset of menstruation (on the 16th day of the cycle of a 30-day cycle) the oestrogen level becomes high enough to stimulate a surge of LH. This stimulates ovulation. Having released the ovum, the primary follicle then forms a corpus luteum and starts to produce progesterone. Under this influence, the endometrial lining is prepared for implantation: glands become convoluted ('secretory phase'). The cervical mucus becomes viscid and hostile to sperm and no longer ferns. If the ovum is not fertilized the corpus luteum breaks down, so hormone levels fall. This causes the spiral arteries in the uterine endothelial lining to constrict and the lining sloughs—hence menstruation.

Menstruation This is the loss of blood and uterine epithelial slough. It lasts 2–7 days and is usually heaviest at the beginning. Normal loss is 20–80mL, median 28mL.

Climacteric The ovaries fail to develop follicles. Without hormonal feedback from the ovary, gonadotrophin levels rise. Periods cease (menopause), usually at ~50 years of age (p 18).

Postponing menstruation (eg on holiday) Try norethisterone 5mg/8h po from 4 days before the period is due until bleeding is acceptable, or take 2 packets of combined contraceptive pills without a break.



Abnormal menstruation

Primary amenorrhoea (see p 12) This is failure to start menstruating. It needs investigation in a 16-year-old, or in a 14-year-old who has no breast development. For normal menstruation to occur she must be structurally normal with a functioning control mechanism (hypothalamic-pituitary-ovarian axis).

Secondary amenorrhoea (see p 12) This is when periods stop for >6 months, other than due to pregnancy. Hypothalamic-pituitary-ovarian axis disorders are common, ovarian and endometrial causes are rare.

Oligomenorrhoea This is infrequent periods. It is common at the extremes of reproductive life when regular ovulation often does not occur. A common cause throughout the reproductive years is polycystic ovary syndrome (p 13).

Menorrhagia (p 14) This is excessive blood loss.

Dysmenorrhoea This is painful periods (\pm nausea or vomiting). 50% of British women complain of moderate pain, 12% of severe disabling pain.

Primary dysmenorrhoea is pain without organ pathology—often starting with anovulatory cycles after the menarche. It is crampy with ache in the back or groin, worse during the first day or two. Excess prostaglandins cause painful uterine contractions, producing ischaemic pain. *Treatment:* Prostaglandin inhibitors, eg mefenamic acid 500mg/8h po during menstruation reduce contractions and hence pain. Paracetamol is a good alternative to NSAIDs. In pain with ovulatory cycles, ovulation suppression with the combined Pill can help (thus dysmenorrhoea may be used as a covert request for contraception). Smooth muscle antispasmodics (eg alverine 60–120mg/8h po) or hyoscine butylbromide (20mg/6h po) give unreliable results. Cervical dilatation in childbirth may relieve it but, surgical dilatation may render the cervix incompetent and is no longer used as therapy.

Secondary dysmenorrhoea: Associated pathology: adenomyosis, endometriosis, chronic sepsis (eg chlamydial infection), fibroids—and so it appears later in life. It is more constant through the period, and may be associated with deep dyspareunia. Treatment of the cause is the best plan. IUCDs increase dysmenorrhoea.

Intermenstrual bleeding This may follow a midcycle fall in oestrogen production. Other causes: cervical polyps; ectropion; carcinoma; cervicitis and vaginitis; hormonal contraception (spotting); iucd; pregnancy-related.

Postcoital bleeding *Causes:* Cervical trauma; polyps; cervical, endometrial and vaginal carcinoma; cervicitis and vaginitis of any cause. Invasive cervical carcinoma may be found in 3.8% of those referred to hospital. Smears may be normal so refer all with persistent bleeding, with a smear with CIN I or worse, wart virus change or atypia¹ (p 34).

Postmenopausal bleeding This is bleeding occurring later than 1yr after the last period. It must be considered due to endometrial carcinoma until proved otherwise (p 40). Other causes: vaginitis (often atrophic); foreign bodies, eg pessaries; carcinoma of cervix or vulva; endometrial or cervical polyps; oestrogen withdrawal (hormone replacement therapy or ovarian tumour). She may confuse urethral, vaginal, and rectal bleeding.

Amenorrhoea

► Always ask yourself 'Could she be pregnant?'

Primary amenorrhoea (see also p 10). This may cause great anxiety. In most patients puberty is just late (often familial), and reassurance is all that is needed. A few causes will be structural or genetic, so check:

- Has she got normal external secondary sexual characteristics? If so, are the internal genitalia normal (p 6)?
- If she is not developing normally, examination and karyotyping may reveal Turner's syndrome (p 759) or testicular feminization (p 222). The aim of treatment is to help the patient to look normal, to function sexually, and, if possible, to enable her to reproduce if she wishes.

Causes of secondary amenorrhoea

- Hypothalamic-pituitary-ovarian causes are very common as control of the menstrual cycle is easily upset, eg by emotions, exams, weight loss, excess prolactin (30% have galactorrhoea), other hormonal imbalances, and severe systemic disease, eg renal failure. Pituitary tumours and necrosis (Sheehan's syndrome) are rare.
- Ovarian causes: polycystic ovary syndrome (opposite), tumours, ovarian failure (premature menopause), are uncommon.
- Uterine causes: pregnancy-related, Asherman's syndrome (uterine adhesions after a D&C). 'Post-Pill amenorrhoea' is generally oligomenorrhoea masked by regular withdrawal bleeds.

12

Tests Serum LH and testosterone (↑ in polycystic ovary syndrome), FSH (very high in premature menopause), prolactin (↑ by stress, prolactinomas and drugs, eg phenothiazines) and TFT are the most useful blood tests. 40% of those with hyperprolactinaemia have a tumour so do MRI scan (p 58).

Treatment is related to cause. Premature ovarian failure cannot be reversed but hormone replacement (p 18) is necessary to control symptoms of oestrogen deficiency and protect against osteoporosis and heart disease. Pregnancy can be achieved with oocyte donation and *in vitro* fertilization techniques.

Hypothalamic-pituitary axis malfunction: If mild (eg stress, moderate weight loss): there is sufficient activity to stimulate enough ovarian oestrogen to produce an endometrium (which will be shed after a progesterone challenge, eg norethisterone 5mg/8h for 7 days), but the timing is disordered so cycles are not initiated. If the disorder is more severe the axis shuts down (eg in severe weight loss). FSH and LH and hence oestrogen levels are low. Reassurance and advice on diet or stress management, or psychiatric help if appropriate (p 348), and time may solve the problem. She should be advised to use contraception as ovulation may occur at any time. If she wants fertility restored now, or the reassurance of seeing a period, mild dysfunction will respond to clomifene (=clomiphene) but a shut-down axis will need stimulation by gonadotrophin-releasing hormone (see p 58 for both).

Polycystic ovarian syndrome (PCOS)^{1,2}

This would be better named polyfollicular ovary syndrome. It comprises hyperandrogenism, oligo-ovulation, and polycystic ovaries on ultrasound in the absence of other causes of polycystic ovaries, eg as seen with later onset adrenal hyperplasia and Cushings. The cause is unknown. It is common (5–20% of premenopausal women). Acne, male pattern baldness and hirsutism can all be features. Darkened skin (acanthosis nigricans) on neck and skin flexures may reflect hyperinsulinaemia.² LH is raised in 40%, testosterone in 30%.² Infertility may be a problem. The result appears to be a vicious circle of ovarian, hypothalamic-pituitary, and adrenal dysfunction.

Insulin resistance and hyperinsulinaemia are features. There may be diabetes (40% of women with the syndrome have impaired glucose tolerance or diabetes by age 40, those of lean habitus also being at greater risk; although the obese are at greatest risk). Insulin resistance is associated with an atherogenic lipid profile predisposing to coronary heart disease, especially in the obese.¹ Stroke and transient ischaemic attack rate is increased (x3).² Hypertension³ may also be a problem; especially affecting older postmenopausal women. Hormonal cycling is disrupted and the ovaries become enlarged by follicles which have failed to rupture. Ovarian cancer risk is increased,⁴ as is endometrial cancer.²

Stein–Leventhal syndrome eponymizes the subset of obese hirsute women with polycystic ovaries.

Management Advise smoking avoidance. Detect and treat diabetes, hypertension, and hyperlipidaemia. Encourage weight loss and increased exercise to increase insulin sensitivity. Metformin may also improve insulin sensitivity, menstrual disturbance, and ovulatory function.⁵

Clomifene (=clomiphene) will usually induce ovulation (50–60% conceive in first 6 months of treatment; offer 12 cycles before proceeding to other methods. Use on specialist advice.) Those who will not ovulate in response to clomiphene are at ↑ risk of ovarian hyperstimulation (p 75) with assisted conception.³ Ovarian drilling (creating holes in ovaries by diathermy with the intent of reducing steroid production) has insufficient information from randomized controlled trials to know if it is effective.²

The combined Pill will control bleeding and reduce risk of unopposed oestrogen on the endometrium (risk of endometrial carcinoma). Some recommend annual ultrasound to check for evidence of endometrial thickening and need for endometrial biopsy.² Regular withdrawal bleeds can be induced with dydrogesterone 10mg/12h po on days 10–25 in those in whom oestrogen use is not wanted or is contraindicated.

Hirsutes may be treated cosmetically, or with an anti-androgen, eg cyproterone 2mg/day, as in Dianette® (avoid pregnancy). Spironolactone 25–100mg/24h/po is also antiandrogenic (avoid pregnancy as teratogenic). Finasteride 5mg/24h/po has also been used for hirsutism,¹ again avoid pregnancy. Neither spironolactone nor finasteride is licensed for this use.

Menorrhagia

This is increased menstrual blood loss (defined as $>80\text{mL/cycle}$); in reality loss is rarely measured, so management deals with those whose significant menstrual loss interferes with their life.¹ What makes a woman consult may be a change in volume (clots; floods, etc), or a change in her life (eg job change, or depression). Ask about both.¹ Is she hypothyroid (eg constipation; weight \uparrow ; *OHCM* p 296) or anaemic? Examine with these in mind.

Causes In girls, pregnancy and dysfunctional uterine bleeding are likely. With increasing age, think also of IUCD, fibroids, endometriosis and adenomyosis, pelvic infection, polyps. In perimenopausal women, consider endometrial carcinoma. Ask about general bleeding problems as she may have a blood dyscrasia, eg von Willebrand's. Do abdominal and pelvic examination¹—which may reveal polyps, fibroids, or endometriosis.

Tests¹ Do FBC; TFT if seems hypothyroid; ultrasound or laparoscopy if pelvic pathology suspected; ultrasound and endometrial sampling, or hysteroscopy and directed biopsy if irregular bleeding or suspected cancer.

Dysfunctional uterine bleeding (DUB) This is heavy and/or irregular bleeding in the absence of recognizable pelvic pathology. It is associated with anovulatory cycles, so is common at the extremes of reproductive life or it may be ovulatory (eg with inadequate luteal phase). If PV is normal and organic pathology is ruled out, this is the diagnosis, by exclusion.

14 Treatment of menorrhagia Treat any underlying condition. For dysfunctional uterine bleeding, treatment depends on age. Reassurance helps. Teenage menorrhagia generally settles without interference as cycles become ovulatory. Those with unacceptable loss should be offered treatment. Refer those with irregular bleeding or bulky uterus (>10 week size).

Drugs *Antifibrinolytics* Taken during bleeding these reduce loss (by 49%)—eg *tranexamic acid* 1g/6–8h po. CI: thromboembolic disease—but this is no more common in those on tranexamic acid. *Antiprostaglandins*, eg *mefenamic acid* 500mg/8h po pc (CI: peptic ulceration) taken during days of bleeding particularly help if there is also dysmenorrhoea. They reduce bleeding by 29%. *Hormones* The *combined Pill* is effective but may be contraindicated in older women—(eg smokers >35 years). *Danazol* 100mg/6–24h po is effective, and expensive. Use only on special advice. It inhibits ovulation (unreliably, so advise condoms as it masculinizes the $\text{\textcircled{f}}$ fetus). Oral cyclical progestogens are not effective.²

Progesterone-containing IUCDs, eg the Mirena[®] are good for those wishing to avoid pregnancy and to reduce bleeding^{~3~} (by up to 86% at 3 months, 97% at 1yr). They are effective in dysfunctional uterine bleeding and should possibly be considered for first-line management of menorrhagia. They have also been shown to reduce fibroid volume after 6–18 months use.

For those women who have completed their families consider endometrial resection or hysterectomy (p 72). Endometrial resection removes the first few millimetres thickness of endometrium; eg by laser or rollerball electrode (safest)³ or by electrocautery loop (most side-effects)⁴ or by heating (microwave, balloon). Ablation is more likely to be effective if the endometrium is thin, and pretreatment with leuproprelin (p 52) may be better than using a progestogen or danazol. About 30% become amenorrhoeic and a further 50% have reduced flow after any method of ablation.

1 M Shapley 2002 *BJGP* 52 108 2 Evidence Based Guidelines 1998 RCOG www.rcog.org.uk/medical/clinical.html ~3~ P Haukka 1999 *E-DM* 4 18 (the need for hysterectomy is \downarrow) 4 C Overton 1998 *Br J Obs Gynaecol* 105 1351

The premenstrual syndrome (PMS)

Most women notice that their mood or physical state may be worse premenstrually. Symptoms may be mild one month and severe the next, eg depending on external events and tend to be worse in the 30s and 40s and improve on the combined Pill. 3% of women regularly have cyclical symptoms so severe that they cause major disruption to their lives: premenstrual syndrome (PMS) or tension (PMT). Causes: See opposite.

Symptoms Commonest symptom patterns are tension and irritability; depression; bloating and breast tenderness; carbohydrate craving and headache; clumsiness; libido↓.¹ Almost any symptom may feature.

Diagnosis Suggest symptom diary. If she has PMS her symptoms are worst before periods, are relieved by menstruation, and there is at least one symptom-free week afterwards. Diaries may also reveal psychiatric disorders (which may be worse premenstrually) or menstrual disorders.

Treatment Simply to acknowledge her problem, listen, and reassure may be all that is needed to enable her to cope. Are her partner and children understanding? Can she rearrange work schedules to reduce stress premenstrually? Some women find self-help groups supportive. Health measures, eg improved diet, reducing smoking and drinking, increased exercise and relaxation, often help. Herbal remedies are not scientifically tested but some find them helpful, eg sage and fennel for irritability. Any drug evokes a big placebo effect, improving 90% in some studies. Pyridoxine (vit B₆) 10mg/24h po for the symptomatic period or continuously, may help low mood and headache (Don says higher, more effective, doses cause neuropathy—but the evidence is flimsy²). For severe *cyclical mastalgia* consider:

- 1 Reduce saturated fats eaten: these increase the affinity of oestrogen-receptors for oestrogen.³
- 2 Gamolenic acid 160mg/12h po. 1 capsule=40mg in evening primrose oil. A contra-indication is a past history of seizures.
- 3 Bromocriptine 2.5mg/12h po days 10–26, even if prolactin normal.
- 4 Danazol 100–200mg/12h po for 7 days before menstruation (see below).

Premenstrual progestogens, eg progesterone suppositories 200mg/12h PR or PV may provide general relief. Some benefit from suppression of ovulation with the combined Pill, oestrogen patches or implants, with cyclical progesterone, or danazol 200mg/24h (se nausea, weight gain, masculinization of the ♀ fetus, so advise barrier contraception). Avoid diuretics unless fluid retention is severe (when spironolactone 25mg/6h po days 18–26 of the cycle is the drug of choice).

For psychological symptoms, fluoxetine 20mg/day po helps, with ~30% experiencing remission. Alprazolam (0.25mg/day po during the luteal phase) also helps, with apparently low risk of dependence. Other treatments for PMT include mefenamic acid (250–500mg/8h po from day 16 until 3 days into period). This may help fatigue, headache, pains, and mood. Agnus castus fruit extract helped these symptoms in 50% of women in a placebo controlled trial.⁴ Goserelin (OHCM p 268) may help severe PMT² but symptoms return when ovarian activity recommences and after 6 months' use bone thinning can be detected. It is better used to predict the severely affected women who may benefit from hysterectomy with oophorectomy (these women can then have oestrogen replacement).

Follow-up: it is not 'cheating' to ensure that her next appointment will not be in the premenstrual phase—more objectivity may be forthcoming.

1 J Butcher 1999 *BMJ* i 41 2 A Beckett 1998 *Lancet* 352 62 3 *Drug Ther Bul* 1992 30 1
4 R Schellenberg 2001 *BMJ* i 134

The Rapkin hypothesis and putative pathways in PMS¹

Some physiological and pharmacological observations:

- There is no evidence that ovarian events cause premenstrual syndrome: models presupposing progesterone deficiency have not been confirmed (and progesterone suppositories are no panacea for the condition).
- Artificially altering circulating progesterone and oestradiol (estradiol) does not induce premenstrual symptoms in previously well women—only in those already prone to PMS.
- Studies with psychoactive compounds suggest that the key events are occurring in the brain, not the ovary—eg an abnormal CNS response to normal progesterone excursions occurring in the luteal phase.
- Allopregnanolone and pregnenolone (metabolites of progesterone) are psychoactive, interacting, eg with γ -aminobutyric acid A (GABA-A) receptors.
- Allopregnanolone is anxiolytic, so low levels may cause anxiety.

Putative conclusion: Neurones or glia in those with PMS preferentially metabolize progesterone to pregnenolone (which heightens anxiety) rather than allopregnanolone (which is anxiolytic and up-regulates serotonin receptors, so ameliorating depression). On this view, alprazolam, by augmenting GABA-A-receptor function, is a substitute for allopregnanolone.

¹ A Rapkin 1997 *Obstet Gynaecol* **90** 709–14 & S Berga 1998 *Lancet* **351** 465

The menopause and HRT

The climacteric is the time of waning fertility leading up to the last period (the menopause). The menopause enables grandmothering—a key institution, so far only discovered by humans and whales. www.txt.co.uk/sum.cfm?1046
Menopause problems are related to falling oestrogen levels:

- Menstrual irregularity as cycles become anovulatory, before stopping.
- Vasomotor disturbance—sweats, palpitations, and flushes (brief but nasty, and may occur every few minutes for >10yrs, disrupting life and sleep).
- Atrophy of oestrogen-dependent tissues (genitalia, breasts) and skin. Vaginal dryness can lead to vaginal and urinary infection, dyspareunia, traumatic bleeding, stress incontinence, and prolapse.
- Osteoporosis. The menopause accelerates bone loss which predisposes to fracture of femur neck, radius, and vertebrae in later life (p 676).
- Arterial disease becomes more common after the menopause.
- Attitudes to the menopause vary widely, and partly depend on irritability, depression, 'empty nest syndrome'—all exacerbated by the menopause.

Management ≥20% of women seek medical help.

- Is it the menopause? Thyroid and psychiatric problems may present similarly. Measure FSH if diagnosis in doubt (very high in menopause).
- Counselling helps psychosocial and physical symptoms. Enlist family's support.
- Menorrhagia may respond to treatment (p 14). A D&C is required if irregular bleeding is abnormal (it may be difficult to decide).
- Continue contraception for 1yr after the last period, eg POP, IUCD, condoms.
- Hot flushes may respond to clonidine 50–75µg/12h PO, HRT, or tibolone.
- Vaginal dryness responds to oestrogen (can be used locally).

Hormone replacement therapy (HRT) Oestrogen is not a panacea for all problems, but may help flushes and atrophic vaginitis. It postpones menopausal bone loss. Randomized trials show no cardiovascular benefits.¹ HRT may prevent Alzheimer's disease (oestrogen is more antioxidant than vit. E, and crosses the blood–brain barrier well, and it ↓ CNS amyloid, and may ↑ cholinergic effects). It reduces proteinuria in diabetic and hypertensive nephropathy.² HRT can ↑ breast cancer risk (below), endometrial cancer, and those using for >10yrs have double the risk of ovarian cancer of non-users.³ As benefits are many; most women should at least consider HRT.

Women with a uterus should also receive progestogens, eg norgestrel 150µg/24h, PO for 12 days out of 28 to reduce risk of endometrial carcinoma; this produces withdrawal bleeds—if unacceptable, consider tibolone 2.5mg/day PO, a preparation which aims not to cause bleeds—or continuous combined oestrogen/progestogen combinations, eg Kliofem®, estradiol (=oestradiol) 2mg and norethisterone 1mg. For both these, either wait for 1yr after the last period, or, if changing from cyclical HRT, wait until after 54yrs of age; bleeding is common, in the 1st 4 months of use—reassure; if after 8 months, do an endometrial biopsy. Kliofem® may be useful if cyclical HRT causes 'premenstrual' symptoms. There may be advantages in using natural 'lipid-friendly' progestogen, dydrogesterone (no androgenic, mineralocorticoid, or oestrogenic action), eg Femoston 2/10® or (2/20 tabs, ie 2mg 17β (o)estradiol tabs, each with added 20mg dydrogesterone after day 14), but see p 66 for possible SE (DVT). Raloxifene, a selective (o)estrogen receptor modulator (SERM) protects bones while reducing breast and endometrial cancer risk. It is ineffective for flushes.⁴

HRT contraindications: • Oestrogen-dependent cancer • Undiagnosed PV bleeding • LFTs↑ • Pregnancy • Breast feeding • Phlebitis • Past pulmonary embolus
Avoid or monitor closely in Dubin–Johnson/Rotor syndromes (OHCM p 722).

BP↑ is not a CI. If past spontaneous DVT/PE: is there thrombophilia (OHCM p 680)?

Side-effects: Weight↑; 'premenstrual' syndrome; cholestasis; vomiting.

Annual check-up: Breasts; BP (stop if BP >160/100 pending investigation and treatment). Weight; any abnormal bleeding?

Creams, pessaries, & rings are useful for vaginal symptoms, eg Ovestin®, ie (o)estriol 0.1%, 1 applicator-full pv daily for 3 weeks, then twice weekly. They are absorbed but, if used intermittently, progestogens are probably unnecessary. If creams are unacceptable, consider an oestrogen-containing vaginal ring (eg Estring®) replaced every 3 months, for up to 2yrs. Vagifem® is an estradiol (=oestradiol) 25µg vaginal tablet daily for 2 weeks, and then twice weekly, with reassessment every 3 months.

Transdermal patches are less 'medical' but are expensive and women with wombs still need progestogen, eg as tablets or Estracombi® patches. Estradiol (=oestradiol) patches supply 25–100µg/24h for 3–4 days. SE: dermatitis.

18(O)estradiol implants (Surgical) 25mg lasts ~36 weeks. They give as good symptom control as 50mg.⁵ Aim for serum oestradiol levels 300–500 pmol/L (300 is sufficient to maintain bone), as levels >500 pmol/L may be associated with dependence.⁶

(O)estradiol gel 2 measures are applied daily to arms, shoulders, or inner thighs. Women with a womb using gel or implants also require progestogens for 12 days per cycle.

When is HRT particularly desirable? ► Make a point of offering HRT to:

- Those who have had premenopausal bilateral oophorectomy.
- Women after hysterectomy (ovaries often fail, eg by 5yrs post-op).
- Those with ↑ risk of osteoporosis (eg smokers or if inactive); see p 676.
- Those with rheumatoid arthritis.

HRT and breast cancer This is the most serious question mark over HRT. If HRT is used for ≥5yrs, relative risk increases by 1.023/yr of HRT use. For this reason our policy on HRT is:

- To discuss the risk of breast cancer with each patient considering HRT.
- To document this discussion in the patient's notes.
- To air issues about breast-awareness during normal activities such as bathing and dressing, and to explain, if necessary, that formal breast examination by nurses may give false reassurance (advice of the UK's chief medical officer¹). The rôle of mammography in detecting HRT-induced breast cancer is unknown. It is difficult if aged 40–50yrs (breast density↑).
- To encourage women to bring up problems with therapy at an early stage.
- Not to prescribe HRT in a blanket fashion just because a woman wants it.
- To be wary about HRT in those with a family history of breast cancer.
- To consider stopping HRT before 6 completed years of therapy.

An alternative for those wanting HRT only for their bones may be raloxifene (which decreases risk of breast cancer: it is related to tamoxifen).⁴

HRT and venous thromboembolism The number of extra cases is only about 1 in 500 users per year. Risk may be concentrated in new users, rather than those who have been taking HRT for many years. The UK committee on safety of medicines (CSM) suggests reviewing need for HRT in those with predisposing factors, such as prolonged immobility, surgery, obesity, past or family history of DVT or embolism, or severe varicose veins.

Termination of pregnancy (TOP)

► Under UK law, no one *has* to have an abortion, and no one *has* to do one. Worldwide, 30% of pregnancies are terminated. Good contraceptive services reduce but do not abolish demand. *Incidence*: ~170,000 TOPs/yr in UK.

Legal (UK) constraints The 1967 and 1990 Human Fertilization/Embryology Acts allow termination before 24 weeks if this reduces the risk:

- 1 To the woman's life, or:
- 2 To her physical or mental health—or:
- 3 Of adverse physical or mental health to her existing children, or:
- 4 Of the baby's being seriously handicapped, mentally or physically.

90% of TOPs are carried out via *Clause 2* ('social'). 2 doctors must sign form HSA 1. Parental consent (and the patient's) is needed for those under 16.

There is no upper time limit if there is: ● Risk to the mother's life.

- Risk of grave, permanent injury to the mother's physical/mental health (allowing for reasonably foreseeable circumstances).
 - Substantial risk that if the child were born he or she would suffer such physical or mental abnormalities as to be seriously handicapped.
- <1% of TOPs are done after 20 weeks, usually after amniocentesis, or when very young or menopausal mothers have concealed, or not recognized, pregnancy. TOPs after 24 weeks may only be carried out in NHS hospitals.

Before TOP ► She has to live with the decision for the rest of her life.

- Counselling to help her reach the decision she will least regret.
- Is she definitely pregnant? Do a VE or scan to confirm dates.
- Is TOP what she really wants? Why? Has she considered alternatives—discuss their implications. What about her partner? Ideally give her time to consider and bring her decision to a further consultation. If she chooses TOP:
- Screen for chlamydia. Untreated, 25% get post-op salpingitis, p 50.
- Routine tetracycline prophylaxis ↓ post-op infections (from 10% to 6%).
- Discuss contraception (eg start Pill next day if she wishes). 90% ♀ ovulate within 1 month of 1st trimester abortion.
- If RhD—ve she needs anti-D (p 109).
- Follow up, eg when the birth would have been (often a fraught time).

Methods *Medical abortion* refers to use of an antigestagen eg mifepristone to disimplant the fetus followed by a prostaglandin eg gemeprost or misoprostol to complete abortion. It is highly effective from ≥6 weeks use (98% effective at ≤7 weeks, 95% weeks 7–9) and is also used for second trimester abortions. Misoprostol can be used orally or vaginally (sometimes both) and is much cheaper than vaginal gemeprost. Gemeprost is more effective when used between 7 and 9 weeks. Arrange follow up (and scan) 2 weeks after procedure unless complete abortion confirmed on the day of abortion; it is for early abortions. 5% will need surgical evacuation.

Dilatation and curettage or vacuum aspiration Used for 1st trimester abortions, women <18 years of age or with gestations >10 weeks require cervical priming pre-operatively. Local anaesthesia can be used (uncommon in UK). Medical termination is preferable if <7 weeks as gestational sacs may be missed so early. Mortality is low (1:100,000), as is infection risk.

Dilatation and evacuation using surgical forceps may be used between 13 and 18 weeks. It is more commonly used in non-NHS establishments. Experienced operators are required. Real time ultrasound facilities reduce uterine perforation rates. After 14 weeks, morbidity and mortality ↑ with gestation.

Complications Infection, later miscarriages, breast cancer (JAMA 1996 275 283) and other imponderables such as coarsening of attitudes to life and death when TOP is done lightly. 'We all dream' of a time when no one needs our services, but for those who do, today, we must show a willingness to help and console.

Some regimens

Early medical abortions:

Mifepristone 600mg po + gemeprost 1mg pv 36–48h later. Observe for 6h post gemeprost as risk of ↓BP; or

*Mifepristone 200mg po + misoprostol 800µg (4 x 200µg tablets) vaginally 36–48h later. (*Unlicensed regimen but given in the guidelines of the Royal College of Obstetricians and Gynaecologists (RCOG).)

Cervical priming:

Gemeprost 1mg pv 3h before surgery; or

*Misoprostol 800 µg (4 x 200µg tablets) pv 3h prior to surgery. (*Unlicensed regimen but given in the guidelines of the RCOG.)

Mid-trimester abortion:

Mifepristone 600mg po followed 36–48h later by gemeprost 1mg pv every 3h to a maximum of 5 pessaries; or

*Mifepristone 200mg po followed 36–48h later by misoprostol 800µg pv; then misoprostol 400µg po every 3h to a maximum of 4 oral doses. (*Unlicensed regimen but given in the guidelines of the RCOG.)

Note: Although 800µg misoprostol is recommended in the guideline, studies show 400µg to be equally effective for cervical dilatation, and this dose can ↓ risk of intra- and post-operative bleeding and abdominal pain.

Recommendations of the 1994–96 Confidential Enquiry into Maternal Mortality

- Ideally all women should undergo ultrasound prior to termination to establish gestational age, viability, and site of pregnancy.
- Failure to obtain tissue at suction termination of a sonographically intrauterine pregnancy suggests abnormal implantation, eg cornual.
- Laparoscopy and/or laparotomy is essential if perforation of the uterus occurs during suction termination to assess any bowel damage.
- Questions remain as to whether dilatation and evacuation is safe and appropriate for second-trimester terminations—when effective medical alternatives exist.

†‘You dream’, he said, ‘because of the child

Asleep in the nest of your body, who dreams. . .’ Stephen Spender, *The Dream*

Abortion (miscarriage)

Abortion is the loss of a pregnancy before 24 weeks' gestation. 20–40% of pregnancies miscarry, mostly in the first trimester. Most present with bleeding *pv*. Diagnosis may not be straightforward (consider ectopics p 24): have a low threshold for doing an ultrasound scan. Pregnancy tests remain +ve for several days after fetal death.

Management of early pregnancy bleeding Consider the following:

- ► Is she shocked? There may be blood loss, or products of conception in the cervical canal (remove them with sponge forceps).
- Has pain and bleeding been worse than a period? Have products of conception been seen? (Clots may be mistaken for products.)
- Is the os open? The external os of a multigravida usually admits a fingertip.
- Is uterine size appropriate for dates?
- Is she bleeding from a cervical lesion and not from the uterus?
- What is her blood group? If RhD–ve does she need anti-D (p 109).

If symptoms are mild and the cervical os is closed it is a *threatened abortion*. Rest is advised but probably does not help. 75% will settle. Threatened abortion (especially second trimester) is associated with risk of subsequent preterm rupture of membranes and preterm delivery—so book mother at a hospital with good neonatal facilities.

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If symptoms are severe and the os is open it is an *inevitable abortion* or, if most of the products have already been passed, an *incomplete abortion*. If bleeding is profuse, consider ergometrine 0.5mg *im*. If there is unacceptable pain or bleeding, or much retained tissue on ultrasound, arrange evacuation of retained products of conception (ERPC). Expectant management is used when volume of retained products is small eg <15mm across on transvaginal scan; when 15–50mm medical management eg with mifepristone may be offered (benefit may not be conclusive).¹

Missed abortion: The fetus dies but is retained. There has usually been bleeding and the uterus is small for dates. Confirm with ultrasound. Mifepristone and misoprostol may be used to induce uterine evacuation if the uterus is small but 50% will require surgical evacuation if uterine products are >5cm² in the transverse plane; >6cm² in the sagittal plane.¹ Surgical evacuation is required for larger uteruses.

Mid-trimester abortion This is usually due to mechanical causes, eg cervical incompetence (rapid, painless delivery of a live fetus), uterine abnormalities; or chronic maternal disease (eg DM, SLE). An incompetent cervix can be strengthened by a cervical encirclage suture at ~16 weeks of pregnancy. It is removed prior to labour.

After a miscarriage ► Miscarriage may be a bereavement. Give the parents space to grieve, and to ask why it happened and if it will happen again.

Most early pregnancy losses are due to aneuploidy and abnormal fetal development; 10% to maternal illness, eg pyrexia. 2nd trimester loss may be due to infection, eg CMV (p 98–100). Bacterial vaginosis has been implicated. Most subsequent pregnancies are normal although at increased risk.

Recurrent miscarriage See opposite.

Septic/'backstreet' abortion Presents as acute salpingitis (p 50) and is treated similarly. Start broad-spectrum antibiotics 1h prior to uterine curettage, eg co-amoxiclav (ampoules are 1.2g; 1g is amoxicillin and 200mg clavulanic acid; give 1.2g/6h *iv*) + metronidazole (eg 1g by suppository/8h).

Causes of recurrent spontaneous miscarriage (RSM)

This is loss of 3 or more consecutive pregnancies. It affects 1% of women.

Possible causes

Endocrine: Polycystic ovaries (found in 58% on ultrasound; the effect is possibly mediated by high luteinizing hormone levels); poorly controlled diabetes; thyroid antibody presence.

Infection: Bacterial vaginosis (p 48) is associated with 2nd trimester loss.

Exposure to chemicals: Smoking, for example, increases abortion rates.¹

Parental chromosome abnormality: ~4% of those with RSM. It is usually a balanced reciprocal or Robertsonian translocation. Refer to a geneticist.

Uterine abnormality: Rates are similar to those in women without RSM so it is probably not a cause.

Antiphospholipid antibodies: (lupus anticoagulant, phospholipid and anticardiolipin antibodies) These are present in 15% of women with RSM. Most women with antibodies miscarry in the first trimester. If they are present, giving aspirin eg 75mg/24h po from the day of positive pregnancy test + heparin (unfractionated self-administered Calciparine®) 5000u/12h sc as soon as the fetal heart is seen (eg at 5 weeks on vaginal ultrasound) until 34 weeks' gestation helps.² Get expert advice.

Alloimmune causes: The theory is that these women share human leukocyte alleles (HLA) with their partners and do not mount the satisfactory protective response to the fetus. Treatment by paternal or third party leukocyte maternal immunization (immunotherapy) is contentious.

Recommended investigations³

- Karyotyping peripheral blood of both parents
- Karyotyping fetal products
- Pelvic ultrasound to assess ovaries and uterus
- Tests for antiphospholipid antibodies twice, taken 6 weeks apart with a third test taken if discordant results.

¹ B Roberta 1999 *NEJM* **340** 333 ² R Rai 1997 *BMJ* **i** 253 Live births with aspirin alone were 42%; with aspirin and heparin 71%. This study is criticized because of the unexpectedly high rate of fetal loss overall; also no cytogenic analysis was done. NB: heparin has serious side-effects; osteoporosis and thrombocytopenia.

³ <http://www.rcog.org.uk/guidelines/recurrent.html>

Ectopic pregnancy

The fertilized ovum implants outside the uterine cavity. The UK incidence is 9:1000 pregnancies and rising; worldwide rates are higher. ~7% of maternal deaths are due to ectopics (1.8 deaths/1000 ectopic pregnancies).

Predisposing factors Anything slowing the ovum's passage to the uterus increases risk: damage to the tubes (salpingitis; previous surgery); previous ectopic; endometriosis; the presence of an older IUCD; the POP (p 66), GIFT (p 58).

Site of implantation 97% are tubal. Most implant in the ampulla, 25% in the narrow inextensible isthmus (so tend to present early and to rupture). 3% implant on the ovary, cervix, or peritoneum.

Natural history The trophoblast invades the tubal wall, weakening it and producing haemorrhage which dislodges the embryo. If the tube does not rupture, the blood and embryo are shed or converted into a tubal mole and absorbed. Rupture can be sudden and catastrophic, or gradual, giving increasing pain and blood loss. Peritoneal pregnancies may survive into the third trimester, and may present with failure to induce labour.

Clinical presentation ▶ Always think of an ectopic in a sexually active woman with abdominal pain or bleeding.

There is generally ~8 weeks' amenorrhoea but an ectopic may present before a period is missed. Tubal colic causes abdominal pain which may precede vaginal bleeding. Blood loss may be dark ('prune juice', as the decidua is lost from the uterus) or fresh. The ectopic may rupture the tube with sudden severe pain, peritonism, and shock. More often there is gradually increasing vaginal bleeding, and bleeding into the peritoneum producing shoulder-tip pain (diaphragmatic irritation) and pain on defecation and passing water (due to pelvic blood). The patient may be faint, with a tender abdomen (95%), enlarged uterus (30%), cervical excitation (50%), adnexal mass (63%). Classical features may be absent. Examine gently, to reduce risk of rupture; preferably with an IVI *in situ*.

Management Early diagnosis is vital. Quantitative β HCG (human chorionic gonadotrophin); do ultrasound. If β HCG >6000iu/L and an intrauterine gestational sac is not seen, ectopic pregnancy is very likely, as is the case if β HCG 1000–1500iu/L and no sac is seen on transvaginal ultrasound. The higher the index of suspicion, the quicker the diagnosis.

▶▶ Shock from a ruptured ectopic can be fatal. *Immediate* laparotomy is necessary as only clamping the bleeding artery will relieve it. If you suspect an ectopic, put up an IVI; if already shocked put up 2 (14 or 16G). Give colloid as fast as possible (use pressure bag to ↑flow), followed by blood—O—ve if desperate, but usually better to wait for group compatible. Inform your consultant. Take immediately to theatre.

A variety of treatments are available in the absence of rupture but surgical treatment remains preferred.¹ Laparoscopy is superior to laparotomy in that recovery time is reduced and rates of subsequent intrauterine pregnancy are higher but persisting trophoblast is more of a problem (12% vs 1.2%). Persistent trophoblast can cause later rupture and will need further treatment (surgical or methotrexate). It is detected by raised β HCG levels. Salpingectomy in the presence of a contralateral healthy tube results in similar rates of subsequent intrauterine pregnancy to salpingotomy (62%) but with lower rates of persisting trophoblast and subsequent ectopic pregnancy. Salpingotomy is preferred if there is only one tube but the subsequent ectopic rate is 20% (inform the patient of this risk). Methotrexate (IM or intratubal injection into the gestation sac) is sometimes used for small (<3.5cm in greatest diameter), early ectopics.

Subsequent fertility rates seem higher after methotrexate treatment.^{2,3} Multiple ovarian cysts, life-threatening neutropenia, pneumonitis, and late pelvic collections of blood have been reported with methotrexate treatment.

Some tubal pregnancies end themselves without any problem so conservative treatment *may* be an option in those without acute symptoms and with falling β HCG levels.

► Remember to give anti-D prophylaxis (p109), if needed.

Measures to reduce missing ectopic pregnancies

- Always send uterine curettings at ERPC (p 72) for histology.
- If histology does not confirm uterine failed pregnancy, recall the patient. (Ensure rapid return of histology results.)
- When ultrasound reports suggest an incomplete abortion but the fetus has not been seen—think: could this be an ectopic?

Gestational [trophoblastic] disease (GTD)

This comprises hydatidiform mole and choriocarcinoma. A fertilized ovum forms abnormal trophoblast tissue, but, usually, no fetus. The growth may appear 'benign' (mole) or 'malignant' (choriocarcinoma)—but these terms are confusing because *normal* trophoblast shows some features of malignancy (invasion and metastasis: in most normal pregnancies trophoblastic tissue ends up, for example, in the mothers lungs—so it may be better to think of the tissue as a failed pregnancy rather than a true neoplasia).

In GTD the trophoblast is usually genetically paternal but has a 46XX karyotype. Rarely, a triploid, partial mole is found with a fetus (usually abnormal). Partial moles do not seem to develop into choriocarcinoma.

Hydatidiform mole The tumour consists of proliferative chorionic villi which have swollen up and degenerated. Since it derives from chorion, it produces human chorionic gonadotrophin (hCG) in large quantities. This gives rise to exaggerated pregnancy symptoms and a strongly +ve pregnancy test. Incidence is 1.54:1000 births (UK). It is more common at extremes of maternal age, after a previous mole, and in non-Caucasians. A woman who has had a previous mole is at increased risk for future pregnancies; 0.8–2.9% after one mole, 15–28% after 2 moles.

Presentation is usually with a pregnancy which is 'not quite right': she may have severe morning sickness or first-trimester pre-eclampsia (20%); the uterus is large for dates in 50% and feels doughy instead of firm. There is no fetal heart. Ultrasound: 'snowstorm' in the uterus.

50% present with vaginal bleeding which is often heavy and prolonged. Molar tissue is likely to be aborted. It looks like a mass of frogspawn.

Abdominal pain may be due to huge theca-lutein cysts in both ovaries. These may rupture or tort. They take ~4 months to resolve after molar evacuation. hCG resembles TSH, and may cause hyperthyroidism. ► Tell the anaesthetist as thyrotoxic storm can occur at evacuation.

Treatment of moles Molar tissue is removed from the soft, easily perforated uterus by gentle suction, followed by a repeat evacuation ~10 days later. Pregnancy should be avoided for a year while hCG levels are monitored. Register the woman at specialist centre (see below) for hCG monitoring. Levels should return to normal within 6 months. If levels drop rapidly to normal, oral contraceptive pills may be used after 6 months. If they do not, either the mole was invasive (myometrium penetrated) or has given rise to choriocarcinoma (10%). Invasive mole may metastasize, eg to lung, vagina, brain, liver, and skin. Both conditions respond to chemotherapy.

Choriocarcinoma This highly malignant tumour occurs in 1:40000 deliveries. 50% follow a benign mole, 20% follow abortions and 10% follow normal pregnancy. *Presentation:* May be many years after pregnancy, with general malaise (due to 'malignancy' and to raised hCG); or uterine bleeding; or with signs and symptoms from metastases, which may be very haemorrhagic, eg haematoperitoneum, or nodules on CXR. Pulmonary artery obstruction via tumour emboli may lead to pulmonary artery hypertension (haemoptysis and dyspnoea).

Treatment: Choriocarcinoma in UK is treated at 3 specialist centres and is extremely responsive to combination chemotherapy based on methotrexate. Outlook is excellent and fertility is usually retained.

► Persistent vaginal bleeding after a pregnancy requires investigation to exclude choriocarcinoma.

The vulva

Pruritus vulvae Vaginal itch is distressing and embarrassing. *Causes:* There may be a disorder causing general pruritus (p 578) or skin disease (eg psoriasis, lichen planus). The cause may be local: infection and vaginal discharge (eg candida); infestation (eg scabies, pubic lice, threadworms); or vulval dystrophy (lichen sclerosis, leukoplakia, carcinoma). Symptoms may be psychogenic in origin. Obesity and incontinence exacerbate symptoms. Postmenopausal atrophy does not cause itch.

The history may suggest the cause. Examine general health and look for widespread skin conditions. Examine the vulva and genital tract, under magnification if possible, and take a cervical smear. Take vaginal and vulval swabs and test for diabetes. Biopsy if in doubt about diagnosis.

►Scratching and self-medication may have changed the appearance.

Treatment: This is often unsatisfactory.¹ Treat the cause if possible. Reassurance can be very important. Advise her to avoid nylon underwear, chemicals, and soap (use aqueous cream) and dry with a hair dryer. A short course of topical steroids, eg betamethasone valerate cream 0.1% may help. Avoid any topical preparation which may sensitize the skin, so give antipruritics orally if needed, eg promethazine 25–50mg/24h.

Lichen sclerosis This is due to elastic tissue turning to collagen after middle age—or, occasionally, before puberty, where the 'bruised' red, purpuric signs appear, to the ignorant, to suggest abuse—particularly if there are bullae, erosions, and ulcerations. The vulva gradually becomes white, flat, and shiny. There may be an hourglass shape around the vulva and anus. It is intensely itchy. It may be pre-malignant and long-term surveillance is desirable.² *Treatment:* Betamethasone valerate cream 0.1% may help; vulval ablation may be needed to relieve itch. In children, 50% resolve by menarche.²

Leukoplakia (White vulval patches due to skin thickening and hypertrophy). It is itchy. It should be biopsied as it may be a pre-malignant lesion.

Treatment: Topical corticosteroids (problems: mucosal thinning, absorption); psoralens with ultraviolet phototherapy; methotrexate; ciclosporin.²

Carcinoma of the vulva 95% are squamous. They are rare and occur mostly in the elderly.

Vulval malignancy has a pre-invasive phase, vulval intra-epithelial neoplasia (VIN), which may be itchy. ~6% progress to invasive carcinoma. VIN is associated with human papilloma virus (HPV) infection. There may not be visible warts but 5% acetic acid stains affected areas white. If VIN is found on biopsy, examine the cervix and check the breasts (>10% have coexistent neoplasia elsewhere, most commonly cervical). *Treatment* is aimed at symptom control by wide local excision³ or laser ablation. Avoid topical steroids³. Recurrence is common so follow up regularly.

An indurated ulcer with an everted edge suggests carcinoma. It may not be noticed unless it causes pain and bleeding, so it often presents late (50% already have inguinal lymph node involvement).

Treatment: Stage 1 and 2 disease (ie tumour <2cm without nodal involvement) may be treated by 'triple incision surgery'. Radical vulvectomy (wide excision of the vulva + removal of inguinal glands) is used if more extensive disease. Post-operative healing may require skin grafts. 5-yr survival is 75% for lesions <2cm without node involvement; otherwise <50%.

¹ Because of the not uncommonly recalcitrant nature of chronic vulval itch and chronic vulval pain (often ignored, or inappropriately diagnosed as candida)—and because of its effects on mood and sexuality, a patients' group (with professional input) has been set up in the UK (Vulval Pain Society, PO Box 514, Slough, Berkshire, SL1 2BP). ² H Bell 1998 *The Diplomat* 5 112 ³ D McConnell 1998 *The Diplomat* 5 118

Vulval lumps and ulcers

Causes of vulval lumps Local varicose veins; boils; sebaceous cysts; keratoacanthomata (rare); viral warts (condylomata acuminata); condylomata lata (syphilis); primary chancre; molluscum contagiosum; Bartholin's cyst or abscess; uterine prolapse or polyp; inguinal hernia; varicocele; carcinoma.

Vulval warts Human papilloma virus (HPV)—is usually spread by sexual contact.) Incubation: weeks. Her partner may not have obvious penile warts. The vulva, perineum, anus, vagina, or cervix may be affected. Wart may be very florid in the pregnant and immunosuppressed. HPV types 16, 18, and 33 can cause vulval and cervical intra-epithelial neoplasia, so she needs annual cervical smears and observation of the vulva. Warts may also cause anal carcinoma (OHCM p 504). Treat both partners. Exclude other genital infections. Warts may be destroyed by diathermy, cryocautery or laser. Vulval and anal warts (condylomata acuminata) may be treated weekly in surgeries and GU clinics with 15% podophyllin paint, washed off after 30min (ci: pregnancy). Only treat a few warts at once, to avoid toxicity. Self-application with 0.15% podophyllotoxin cream (Warticon® 5g tubes—enough for 4 treatment courses—is supplied with a mirror): use every 12h for 3 days, repeated up to 4 times at weekly intervals if the area covered is <4cm². Relapse is common. NB: HPV types 6 and 11 may cause laryngeal or respiratory papillomas in the offspring of affected mothers (risk 1:50–1:1500; 50% present at <5yrs old).

Exclude other genital infections. Treat both partners.

Urethral caruncle This is a small red swelling at the urethral orifice. It is caused by meatal prolapse. It may be tender and give pain on micturition. *Treatment:* Excision or diathermy.

Bartholin's cyst and abscess The Bartholin's glands and their ducts lie under the labia minora. They secrete thin lubricating mucus during sexual excitation. If the duct becomes blocked a painless cyst forms; if this becomes infected the resulting abscess is extremely painful (she can not sit down) and a hugely swollen, hot red labium is seen. *Treatment:* The abscess should be incised, and permanent drainage ensured by marsupialization, ie the inner cyst wall is folded back and stitched to the skin. *Tests:* Exclude gonococcal infection.

Vulvitis Vulval inflammation may be due to infections, eg candida (p 48), herpes simplex; chemicals (bubble-baths, detergents). It is often associated with, or may be due to, vaginal discharge.

Causes of vulval ulcers: Always consider syphilis. *Herpes simplex* is common in the young. Others: carcinoma; chancroid; lymphogranuloma venereum; granuloma inguinale; TB; Behçet's syndrome; aphthous ulcers; Crohn's.

Herpes simplex Herpes type II, sexually acquired, classically causes genital infection, but type I transferred from cold sores can be the cause. The vulva is ulcerated and exquisitely painful. Urinary retention may occur. *Treatment:* Strong analgesia, lidocaine (=lignocaine) gel 2%, salt baths (and mic-turating in the bath) help. Exclude coexistent infections. Aciclovir topically and 200mg 5 times daily po for 5 days shortens symptoms and infectivity. Reassure that subsequent attacks are shorter and less painful. Prescribe aciclovir cream for use when symptoms start. For herpes in pregnancy, see p 100.

The cervix

This is the part of the uterus below the internal os. The endocervical canal is lined with mucous columnar epithelium, the vaginal cervix with squamous epithelium. The transition zone between them—the squamo-columnar junction—is the area which is predisposed to malignant change.

Cervical ectropion This is often called erosion, an alarming term for a normal phenomenon. There is a red ring around the os because the endocervical epithelium has extended its territory over the paler epithelium of the ectocervix. Ectropions extend temporarily under hormonal influence during puberty, with the combined Pill, and during pregnancy. As columnar epithelium is soft and glandular, ectropion is prone to bleeding, to excess mucus production, and to infection. *Treatment:* Cautery will treat these if they are a nuisance; otherwise no treatment is required.

Nabothian cysts These mucus retention cysts found on the cervix are harmless. *Treatment:* Cautery if they are discharging.

Cervical polyps These pedunculated benign tumours of endocervical epithelium may cause increased mucus discharge or postcoital bleeding. *Treatment:* In young women they may be simply avulsed, but in older women treatment usually includes D&C to exclude intrauterine pathology.

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Cervicitis This may be follicular or mucopurulent, presenting with discharge. *Causes:* Chlamydia (up to 50%), gonococci, or herpes (look for vesicles). Chronic cervicitis is usually a mixed infection and may respond to antibacterial cream. Cervicitis may mask neoplasia on a smear.

Cervical screening Cervical cancer has a pre-invasive phase: cervical intra-epithelial neoplasia (CIN—not to be pronounced 'sin'). Papanicolaou smears collect cervical cells for microscopy for dyskaryosis (abnormalities which reflect CIN). A smear therefore identifies women who need cervical biopsy. The degree of dyskaryosis approximates to the severity of CIN (Table, p 35). ~50% of CIN I lesions return to normal but most CIN III lesions progress to invasive carcinoma. This may take ~10yrs, but may happen much faster in young women.

A programme of 2 smears in the first year of sexual activity, then 3-yearly smears should reduce the incidence of invasive carcinoma by 90%. The trouble is, those most at risk are the hardest to trace and persuade to have screening, eg older women, smokers, and those in inner cities. One of the great achievements of UK primary care and its software houses is that these women are now being reached: 83% of the eligible UK population is now screened, a figure which no other country exceeds, and mortality here is starting to fall; this depends critically on being able to retain skilled lab staff.

Taking a smear Explain to the woman the nature and purpose of the test, and how results will be conveyed to her. It may be sensible to warn that results are not categoric or unequivocal.

The cervix is visualized with a speculum (p 2). Are there any suspicious areas? If so, carry on with the smear and indicate this on the referral form, but do not wait for its results before arranging further care.

Cells are scraped from the squamo-columnar transformation zone with a special spatula or brush, then transferred to a slide and fixed at once. Good technique is needed (make sure that all 4 quadrants of the cervix are sampled); it is best to learn by instruction from an expert at the bedside.

Cervical carcinoma

► **Aim to detect pre-invasive disease.** ~1600 women die yearly of cervical cancer in the UK. Many have had no smear, but this is changing: see p 32.

Causes The main cause is human papilloma virus (eg HPV 16, 18, and 33). Prolonged Pill use is thought to be an important co-factor (may ↑ risk 4-fold in those +ve for HPV DNA).¹ Other risk factors: high parity; many sexual partners or a partner having many other partners (esp. if that male is uncircumcized);² early first coitus; HIV; other sexually transmitted diseases; smoking

Management of abnormal smears (See p 35: the histology of cervical intra-epithelial neoplasia (CIN) is explained there.) Either a repeat smear or colposcopy and biopsy are needed, depending on likelihood of the smear reflecting CIN III or small volume invasive disease (<3mm).

► Abnormal smears cause anxiety and guilt. Explain. Give support.

Treatment of pre-invasive carcinoma Examine the cervix by colposcope (×10 binocular microscope). Abnormal epithelium has characteristic blood vessel patterns and stains white with acetic acid. Take punch biopsies for histology. CIN is destroyed by cryotherapy, laser, cold coagulation, or large loop excision of transformation zone (LLETZ). These give ~90% cure rates with one treatment. She needs annual smears for at least 5 years. If the squamo-columnar junction cannot be seen, or if small volume invasive carcinoma is found on histology, the abnormal tissue is removed by cone biopsy, which may be curative. Colposcopy does not detect adenocarcinoma (it usually lies within endocervical canal).

Invasive disease Most are squamous cancers. 15–30% are adenocarcinomas (from endocervical epithelium, with unknown risk factors) especially affecting women under 40. Spread is local and lymphatic. Stage I tumours are confined to the cervix.

Stage II have extended locally.

Stage III have spread to pelvic wall and lower 1/3 of vagina.

Stage IV have spread to bladder or rectum. Most present in stages I or II.

Diagnosis ► Overt carcinoma is rarely detected on a smear. Non-menstrual bleeding is the classic symptom. The early tumour is firm. It grows as a friable mass which bleeds on contact.

Treating invasive cancers Wertheim's hysterectomy or radiotherapy treat the tumour and lymph nodes, and achieve good cure rates at stages I (80% 5-yr survival) and II (60%). Radiotherapy causes vaginal stenosis: so encourage intercourse within 2 months of treatment (+ lubricant). Chemoradiation is the 'gold standard' for stage 1b, 2 or bulky stage 2 disease. Follow-up: annual smears. Smears are of no value after radical radiotherapy. Stage III needs extensive radiotherapy and chemotherapy, eg 5-FU + cisplatin; exenteration is probably *not* indicated (p 73). Stage IV: ?palliation only. Terminal problems are pain, fistulae, and GI/GU obstruction.

1 IARC multicentric cervical cancer study group 2002 *Lancet* 359 1085 and 1080 (editorial)
2 X Bosch 2002 *NEJM* 346 1005; n=1913; occult ♂ HPV infection is common [2556893]

Histology of cervical pre-malignant disease (CIN = cervical intra-epithelial neoplasia)

	<i>Papanicolaou class</i>	<i>Action</i>	<i>Histology</i>
I	Normal	Repeat in 3 years (Unless clinical suspicion)	0.1% CIN II–III
II	Inflammatory	Take swab; treat infection Repeat at 6 months (colposcope after 3 abnormal)	6% CIN II–III
	Mild atypia	Repeat in 4 months (Colposcopy after 2 abnormal)	20–37% CIN II–III
III	Mild dyskaryosis	Colposcopy	20–30% CIN II–III
	Moderate dyskaryosis	Colposcopy	50–75% CIN II–III
IV	Severe dyskaryosis	Colposcopy	80–90% CIN II–III
	'Positive'		5% Invasion
	'Malignant cells'		
V	Invasion suspected	Urgent colposcopy	50% Invasion
	Abnormal glandular cells	Urgent colposcopy	?Adenoca cervix or endometrial ca

CIN I = mild dysplasia; CIN II = moderate dysplasia; CIN III = Severe dysplasia/carcinoma-in-situ

The table above shows comparative terms and the recommended action. The third column, headed histology, shows the percentage of smears in each Papanicolaou (cytological) class which have more serious lesions (CIN II or III) on histology. With inflammatory smears, swabs should be taken and any infection treated. 6% of inflammatory smears have serious pathology, hence the recommendation for colposcopy if inflammation persists.

Borderline nuclear abnormality (BNA) implies doubt as to the neoplastic nature of any change. Re-smearing (eg at 3–6 months), or colposcopy (eg after 2 smears with BNA, if numbers of affected cells are static over time). Such colposcopy should be seen as part of the screening process, and need not prompt treatment. This goes part of the way to avoid false negatives, as well as to avoid the problems of overtreatment.

In the UK, 6×10^6 smears are done per year on women up to 65yrs old, and cervical cancer deaths have fallen by 15% recent years to <1900/yr. 2.4% of smears show mild dyskaryosis, and 2.2% have BNA. As most of these will eventually need colposcopy because of further smear results and 48% can be expected to have CIN II or III, and as a significant proportion may be lost to follow-up, it is suggested that *immediate* colposcopy with directed biopsy is indicated. However,⁶ this would be very expensive, and anxiety-provoking. The cost in the USA of adopting this policy is ~\$1 billion, and the benefits are uncertain as no randomized trials have been done.

Terminology used in reporting smears changed in the 1980s, and different countries use different nomenclature—eg the Bethesda system.

The uterus

Endometritis Uterine infection is uncommon unless the barrier to ascending infection (acid vaginal pH and cervical mucus) is broken, eg after abortion and childbirth, IUCD insertion, or surgery. Infection may involve Fallopian tubes and ovaries.

Presentation: Lower abdominal pain and fever; uterine tenderness on bimanual palpation. Low-grade infection is often due to chlamydia. *Tests:* Do cervical swabs and blood cultures. *Treatment:* Give antibiotics (eg doxycycline 100mg/12h po with metronidazole 500mg/8h po, eg for 7 days).

Endometrial proliferation Oestrogen stimulates endometrial proliferation in the first half of the menstrual cycle; it is then influenced by progesterone and is shed at menstruation. A particularly exuberant proliferation is associated with heavy menstrual bleeding and polyps.

Continuous high oestrogen levels (eg anovulatory cycles) make the endometrium hyperplastic ('cystic glandular hyperplasia'—a histological diagnosis after D&C). It eventually breaks down, causing irregular bleeding (dysfunctional uterine bleeding). *Treatment:* Cyclical progestogens (p 14).

In older women proliferation may contain foci of atypical cells which may lead to endometrial carcinoma (p 40).

Pyometra This is a uterus distended by pus eg associated with salpingitis or secondary to outflow blockage. *Treatment:* Drain the uterus, treat the cause.

Haematometra This is a uterus filled with blood due to outflow obstruction. It is rare. The blockage may be an imperforate hymen in the young (p 6); carcinoma; or iatrogenic cervical stenosis, eg after cone biopsy.

Endometrial tuberculosis Genital tract tuberculosis is rare in Britain, except among high-risk groups (eg immigrants). It is blood-borne and usually affects first the Fallopian tubes, then the endometrium.

It may present with acute salpingitis if disease is very active, or with infertility, pelvic pain, and menstrual disorders (40%) eg amenorrhoea, oligomenorrhoea. There may be pyosalpinx. Exclude lung disease by CXR.

Treatment is medical with antituberculous therapy (OHCM p 566–568). Repeat endometrial histology after one year. Total abdominal hysterectomy with bilateral salpingo-oophorectomy is treatment of choice if there are adnexal masses and the woman is > 40yrs.¹

Uterine ultrasound² Transvaginal ultrasound gives better resolution than transabdominal (as the probe is closer to the target and a higher frequency transducer can be used). Homogeneity, echoes of low intensity and presence of a linear central shadow are associated with absence of endometrial abnormality. Endometrial carcinoma is suggested by endometrial thickness >20mm, heterogeneous appearance, and hypoechoic areas. Polyps have cystic appearance (also with hyperechoic endometrium).

If postmenopausal and not on HRT, double-layer endometrial thickness should be <5mm (if perimenopausal <5mm on day 5 of cycle). Sequential hormone replacement ↑endometrial thickness (average 5–8.5mm); if on continuous combined replacement HRT thicknesses are ~4.5–7mm; tibolone treated endometrium <5mm; but tamoxifen thickens it to ~13mm. It thins down by 6 months after stopping tamoxifen, then stays thin.³

Ultrasound is useful for detecting fibroids; and assessing cystic change in rapidly growing fibroids to assess risk of malignant change.

Vaginal carcinoma

These tumours are usually squamous. They are commonest in the upper third of the vagina. Presentation is usually with bleeding. Clear cell adenocarcinoma was thought to be associated with intrauterine exposure to diethylstilboestrol but risk is low (0.1–1:1000). Spread is local and by lymphatics. Treatment is usually radiotherapy.

Fibroids (uterine leiomyomata)

Fibroids are benign smooth muscle tumours of the uterus (leiomyomas). They are often multiple, and vary in size from seedling size to tumours occupying a large part of the abdomen. They start as lumps in the wall of the uterus but may grow to bulge out of the wall so that they lie under the peritoneum (subserosal, 20%) or under the endometrium (submucosal, 5%), or become pedunculated. Fibroids are common (20% of women have fibroids), increasing in frequency with age, and in non-Caucasians.

Associations Mutation in the gene for fumarate hydratase can cause fibroids and a rare association with skin & uterine leiomyomata, and renal cell cancer.¹

Natural history Fibroids are oestrogen-dependent. Consequently they enlarge in pregnancy and on the combined Pill and atrophy after the menopause. They may degenerate gradually or suddenly (red degeneration). Occasionally they calcify ('womb stones'). Rarely, they undergo sarcomatous change—usually causing pain, malaise, bleeding, and increase in size in a postmenopausal woman.

Presentation Many are asymptomatic.

- **Menorrhagia:** Fibroids often produce heavy and prolonged periods. They do not generally cause intermenstrual or postmenopausal bleeding.
- **Fertility problems:** Submucosal fibroids may interfere with implantation ('natural IUCD'). Large or multiple tumours which distort the uterine cavity may cause abortion should pregnancy occur.
- **Pain:** This may be due to torsion of a pedunculated fibroid, producing symptoms similar to that of a torsed ovarian cyst. 'Red degeneration' following thrombosis of the fibroid blood supply: see page 39.
- **Mass:** Large fibroids may be felt abdominally. They may press on the bladder, causing frequency, or on the veins, causing oedematous legs and varicose veins. Pelvic fibroids may obstruct labour or cause retention of urine.

Treatment In many women treatment is not needed. *Menorrhagia* due to fibroids tends to respond poorly to anti-prostaglandins, progestogens, or danazol. Women who have completed their families may opt for hysterectomy. In younger women a reversible menopausal state may be induced with analogues of LHRH (leutinizing hormone releasing hormone), e.g. buserelin nasal spray 100µg/4h (expensive)¹ or goserelin 3.6mg by monthly subcutaneous injection (preferred method—intermittent buserelin use can increase their size rather than shrink them). Bone demineralization can occur with their use, so their place is in shrinking fibroids prior to surgery (to reduce blood loss at surgery and make surgery easier) in those unfit for surgery; or those desiring later pregnancy. They reduce bulk by 40% over 12 weeks, but no further reduction occurs after 24 weeks use. Side-effects are menopausal symptoms; fertility (and fibroids) return when the drug is stopped. Alternatively, fibroids may be surgically shelled out (myomectomy); torrential bleeding necessitating hysterectomy is a complication and post-operative adhesions notorious. Laparoscopic surgery with or without use of lasers is possible but requires considerable patience. Embolization of fibroids as an interventional radiological procedure can shrink fibroids so resolving menorrhagia, involving only a short hospital stay: but it is not widely available. *Red degeneration* requires only analgesia until symptoms settle. *Torsion* may resemble an acute abdomen requiring urgent surgery.

11 Tomlinson 2002 *Nature Genetics* 2.25 [@:11865300] Gene location: 1q42.3--q43

Fibroids in pregnancy

5/1000 Caucasian women have fibroids in pregnancy. They are commoner in Afro-Caribbean women. They may cause abortion. They increase in size in pregnancy—especially in the second trimester. If pedunculated they may tort. Red degeneration is when thrombosis of capsular vessels is followed by venous engorgement and inflammation, causing abdominal pain (\pm vomiting and low grade fever), and localized peritoneal tenderness—usually in the second half of pregnancy or the puerperium. Ultrasound aids diagnosis. 'Here, as a certain feverishness leads them to their final degeneration', and imitating the course of all grand passions, 'they grow big and tender, and then die' (DH Lawrence, *Sons & Lovers*, JPS, 324).

Treatment is expectant (bed rest, analgesia) with resolution over 4–7 days.

Most fibroids arise from the body of the uterus and do not therefore obstruct labour, as they tend to rise away from the pelvis throughout pregnancy. If large pelvic masses of fibroids are noted prior to labour Caesarean section should be planned. Obstruction of labour also needs Caesarean section.

Endometrial carcinoma

► Postmenopausal vaginal bleeding must be investigated as the cause may be endometrial carcinoma.

Carcinoma of the uterine body is less common than carcinoma of the cervix. It usually presents after the menopause. Most tumours are adenocarcinomas and are related to excessive exposure to oestrogen unopposed by progesterone. There is marked geographical variation: North American: Chinese ratio = 7:1.

Risk factors

- Obesity
- Nulliparity
- Diabetes
- Unopposed oestrogen therapy
- Late menopause
- Pelvic irradiation
- Functioning ovarian tumour
- Tamoxifen*
- FH of breast, ovary, colon cancer
- Polycystic ovaries¹

Presentation This is usually as postmenopausal bleeding (PMB). Any woman with a history of PMB has a 10–20% risk of genital cancer. It is initially scanty and occasional, perhaps with a watery discharge. Over time, bleeding becomes heavier and more frequent. Pre-menopausal women may have intermenstrual bleeding, but 30% have only menorrhagia.

Diagnosis Postmenopausal bleeding is an early sign, and generally leads a woman to see her doctor, but examination is usually normal. Endometrial carcinoma can sometimes be seen on a smear. Uterine ultrasound may be suggestive (p 36). The diagnosis is made by uterine sampling (p 41) or curettage. All parts of the uterine cavity must be sampled; send *all* material for histology. Hysteroscopy enables visualization of abnormal endometrium to improve accuracy of sampling. Sceptics claim it may cause spread through the Fallopian tubes to the peritoneum.

Pathology Most tumours start in the fundus of the uterine cavity. The tumour spreads slowly to the uterine muscle. With time, it may reach the cervix or peritoneum and may metastasize to the vagina (5%), ovary (5%), and any of the pelvic lymph nodes (7%).

Staging

- I: Tumour is confined to the body of the uterus.
- II: As for I but with cervical involvement.
- III: Tumour has extended beyond the uterus but not beyond the pelvis.
- IV: Extension outside the pelvis. Bowel and bladder may be affected.

Treatment Stages I and II may be cured by total hysterectomy with bilateral salpingo-oophorectomy and/or radiotherapy if unfit for surgery (5-yr survival: stage I = 72%, stage II = 56%). Post-operative vault irradiation reduces vault recurrence. In advanced disease consider radiotherapy and/or high-dose progestogens, eg medroxyprogesterone acetate 250mg/24h po, which shrinks the tumour (se: fluid retention). Radiotherapy may either be given pre-operatively (caesium or radium rods inserted into the uterus and upper vagina) or post-operatively (external radiation).

Recurrent disease usually presents in the first 2–3yrs. Common sites are pelvic (in nonirradiated patients), lung, bone, inguinal, and supraclavicular nodes, vagina, liver, peritoneal cavity. Surgical exenteration, radiotherapy, and medroxyprogesterone may all be of use. Cytotoxics may be used for those who fail to respond to the above.

*Note: tamoxifen 20mg daily reduces annual risk of breast cancer recurrence by 27%. Risk of endometrial carcinoma from taking tamoxifen is 1.2 per 1000 person-years. Counsel to report abnormal vaginal bleeding.

Endometrial sampling in outpatients

This bedside investigation is used for postmenopausal bleeding, perimenopausal irregular bleeding, and unexpected bleeding patterns in women on hormone replacement therapy because it is cheap, reliable, and gives quick results without the need for anaesthesia. If transvaginal uterine ultrasound precedes the procedure, sample if endometrium $>5\text{mm}$ thick. It is less useful in menorrhagia in women with regular cycles, as pathology is less common. It is not indicated if <35 years old.

A sample is obtained using a side-opening plastic cannula in which a vacuum is created by withdrawal of a stopped plunger mechanism. As the cannula is then withdrawn and rotated within each quadrant of the uterine cavity, endometrial tissue is sucked into its interior, through the hole in the side of the cannula. Successful insertion is possible in 90–99% of women (p&c possible in 99%). Adequate samples will be obtained in 91% of these, and in 84% of those for whom postmenopausal bleeding (PMB) was the indication. Abandon the procedure if it is impossible to enter the uterus, or if it causes too much pain.

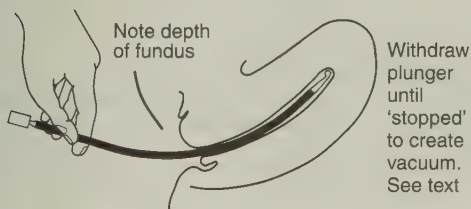
Technique

- 1 Bimanual examination to assess size and position of uterus (p 2).
- 2 Bend cervical cannula to follow the curve of the uterus.
- 3 Insert device, watching the centimetre scale on the side; observe resistance on entering the internal os (at 3–4 cm) and then as the tip reaches the fundus (eg at 6cm if postmenopausal or 8cm in an oestrogenized uterus).
- 4 When the tip is in the fundus, create a vacuum by withdrawing plunger until the stopper prevents further withdrawal. Then move sampler up and down in the uterus, rotate and repeat to sample whole cavity.
- 5 Remove cannula, and expel tissue into formalin. Send for histology.

Vabra vacuum aspiration samples a greater area of tissue, has higher cancer detection rates, but is more uncomfortable.

medline:10428515

Management Reassure those in whom the results show normal or atrophic endometrium and those in whom tissue was sufficient for diagnosis. If those with PMB re-bleed refer for hysteroscopy (polyps or a fibroid will be present in 20%). Those with simple hyperplasia on histology can be treated with cyclical progestones (but refer if $>55\text{yrs}$ to search for exogenous oestrogen source). Refer those with polyps or necrotic tissue on histology for hysteroscopy and curettage; and those with atypical hyperplasia or carcinoma for hysterectomy and bilateral salpingo-oophorectomy. If transvaginal uterine ultrasound not already done, perform in those on whom the procedure was impossible or abandoned to establish endometrial thickness ($<5\text{mm}$ normal in the postmenopausal; refer if $>5\text{mm}$ thick or if polyps seen, for hysteroscopy and curettage).



We thank Genesis Medical Ltd for permission to reproduce the diagram

Ovarian tumours

Any of the ovary's many tissue types may become neoplastic.

Benign tumours (94%). They are usually cystic. 24% of all ovarian tumours are functional cysts. Others: endometriotic cysts (5%—p 52); theca-lutein cysts (p 26); epithelial cell tumours (serous and mucinous cystadenomas—40%); mature teratomas (from germ cells—20%); fibromas (solid—5%).

Malignant tumours (6%). 5% are cystadenomas which have become malignant. 0.5% are a group of rare germ cell or sex cord malignancies (p 43). 0.5% are secondaries, eg from the uterus, or the stomach (Krukenberg tumours—in which spread is transcoelomic, ie, in the case of the abdomen, via the peritoneum).

Risk markers of ovarian malignancy: Nulliparity (risk $\uparrow \times 1.5$); infertility; early menarche; +ve family history (p 46); no past use of the Pill; high total fat intake (associated with a 24% \uparrow risk);¹ ovulation-inducing treatments of infertility.^{6,2}

Presentations are varied, depending on size, form, and histological type: *Asymptomatic*—chance finding (eg on doing a bimanual for a smear test).

Swollen abdomen—with palpable mass arising out of the pelvis which is dull to percussion (and does not disappear if the bladder is catheterized).

42 *Pressure effects* (eg on bladder, causing urinary frequency).

Infarction/haemorrhage—this mimics torsion (see below).

Rupture \pm local peritonism. Rupture of a large cyst may cause peritonitis and shock. Rupture of a malignant cyst may disseminate malignant cells throughout the abdomen. Rupture of mucinous cystadenomas may disseminate cells which continue to secrete mucin and cause death by binding up the viscera (pseudomyxoma peritonei). See p 43. Pseudomyxoma peritonei is treated by surgical debulking. 10-year survival is 30–50%.

Ascites—shifting dullness suggests malignancy or Meigs' syndrome (p 43). If tense, ascites may be hard to distinguish from a mass.

Torsion—to twist, a tumour must be small, and free on a pedicle. Twisting occludes the venous return but the arterial supply continues to engorge the tumour, and cause great pain (with a high WBC). Tumours may twist and untwist, giving a history of intermittent pain. If the pain is not too severe, a firm tender adnexal swelling may be felt.

Endocrine or metastatic effects—Hormone-secreting tumours may cause virilization, menstrual irregularities, or postmenopausal bleeding.

Management *Ultrasound* may confirm the presence of a mass and may show whether it is cystic or solid.

Laparoscopy may distinguish a cyst from an ectopic pregnancy or appendicitis. Note: laparoscopy is not advised if malignancy is possible, due to seeding along the surgical tract. *Fine needle aspiration* may be used to confirm the impression that a cyst is benign. Urgent *laparotomy* is required when a cyst problem presents as an acute abdomen.

Any cyst not positively identified as non-neoplastic should be removed, as seemingly benign tumours may be malignant. In younger women *cystectomy* may be preferable to oophorectomy. In postmenopausal women, if one ovary is pathological both ovaries and the uterus are removed. For management in pregnancy see p 44.

Ovarian tumours: pathology

Functional cysts These are enlarged or persistent follicular or corpus luteum cysts. They are so common that they may be considered normal if they are small (<5cm). They may cause pain by rupture, failing to rupture at ovulation, or bleeding.

Serous cystadenomas These develop papillary growths that may be so prolific that the cyst appears solid. They are commonest in women aged 30 to 40 years. About 30% are bilateral and about 30% are malignant.

Mucinous cystadenomas The commonest large ovarian tumours; these may become enormous. They are filled with mucinous material and rupture may rarely cause pseudomyxoma peritonei (p 42). They may be multilocular. They are commonest in the 30–50 age group. About 5% will be malignant. Remove the appendix at operation in those with suspected mucinous cystadenoma and send for histology.¹ (Interestingly men can get pseudomyxoma from intestinal or appendicular neoplasms; most women with pseudomyxoma peritonei do not have overt rupture of ovarian tumours and 90% have concurrent intestinal or appendicular tumours and it is now thought that the ovarian tumours may be secondary to GI ones).

Fibromas These are small, solid, benign, fibrous tissue tumours. They are associated with Meigs' syndrome (OHCM p 730) and ascites.

Teratomas These arise from primitive germ cells. A benign mature teratoma (dermoid cyst) may contain well differentiated tissue, eg hair, teeth. 20% are bilateral. They are most common in young women. Poorly differentiated malignant teratomas are rare.

Other germ cell tumours (all malignant and all rare) Nongestational choriocarcinomas (secrete HCG); ectodermal sinus tumours (yolk sac tumours—secrete α -fetoprotein); dysgerminomas.

Sex-cord tumours (rare; usually of low-grade malignancy) These arise from cortical mesenchyme. Granulosa-cell and theca-cell tumours produce oestrogen and may present with precocious puberty, menstrual problems, or postmenopausal bleeding. Arrhenoblastomas secrete androgens.

¹ N Sattar 1998 *Lancet* 351 305

Ovarian tumours in pregnancy

These are found in $\sim 1/1000$ pregnancies. It is easier to distinguish them (lying as they do in the rectovaginal pouch) with an anteverted uterus than with a gravid retroverted uterus. Suspicion of presence of a tumour can be confirmed by ultrasound. Torsion of ovarian cysts is more common in pregnancy and the puerperium than at other times. Cyst rupture and haemorrhage into cysts may also occur, but are not more common than at other times. Torsion may present with abdominal pain, nausea, vomiting, shock, local tenderness (usually at 8–16 weeks). 2–5% of tumours are malignant. Suspect malignancy with ruptures (then biopsy other ovary). $\sim 25\%$ of malignant tumours will be dysgerminomas.

Tumours can become necrotic due to pressure on them in labour. Tumours lying in the pelvis can obstruct labour so Caesarean section will be needed unless they are cysts which can be aspirated under ultrasound control before labour.

Asymptomatic simple cysts $< 5\text{cm}$ across can be left until after delivery if watched by ultrasound. Those 5–10cm may be aspirated under ultrasound control (and the aspirate examined cytologically). Other tumours (those that are complex multilocular or with solid portions on ultrasound) should be removed at about 16 weeks' gestation (by which time the pregnancy is not dependent on the corpus luteum and miscarriage is less likely) to exclude carcinoma and prevent complications developing. If the diagnosis is made late in pregnancy and the tumour is not obstructing the pelvis, it is usual to let labour progress normally, and to remove the tumour in the early puerperium because of the risk of torsion then.

Ovarian carcinoma

This is rare, but more women die from it (it is the fourth commonest cause of cancer-related death in Western women) than from carcinoma of the cervix and uterine body combined because in 75% it causes few symptoms until it has metastasized, often to the pelvis with omental and peritoneal seedlings (\pm lymphatic spread via the para-aortic nodes).

Incidence 1 in 2500 women >55 yrs; 1 in 3800 if >25 yrs. Women with 2 close relatives affected have a 40% risk of ovarian cancer (liaise with a gynaecologist; see below). 10% of affected North American women carry mutations in *BRA1* or *BRA2* genes. Carrier risk of developing carcinoma is 40% for women with *BRA1* mutation, 25% if *BRA2*. It is commoner in those with many ovulations (early menarche, nullipara) and, possibly, after infertility treatment (p 58). Combined oral contraceptive Pill use reduces risk.

Presentation Symptoms are often vague and insidious and include abdominal pain, discomfort, and distension.

Screening There is really no good screening test. Transvaginal ultrasound with tumour blood flow measurement can differentiate between benign and malignant neoplasms at an early stage. Plasma levels of cancer associated antigen (CA-125) is insufficiently sensitive or specific.

Consider prophylactic oophorectomy when older women have hysterectomy. It is used for those with *BRA1* & 2 mutations. The combined oral contraceptive Pill reduces the risk (up to 40%). Tubal ligation also reduces risk in those with *BRA1* mutation.¹

Diagnosis Histology. Ascites, ultrasound/CT, and CA-125 \uparrow suggestive.

Staging at laparotomy

Stage I: Disease limited to 1 or both ovaries.

Stage II: Growth extends beyond the ovaries but confined to the pelvis.

Stage III: Growth involving ovary and peritoneal implants outside pelvis (eg superficial liver), or +ve retroperitoneal or inguinal nodes.

Stage IV: Those with distant metastases (including liver parenchyma).

80% present with stage III or IV disease. 5-yr survival: Stage I, 67%; Stage II, 42%; Stage III and IV, 14.4%.

Treatment This depends on the type of tumour. Cystadenocarcinomas (80%) are treated with surgery and chemotherapy, which aims for cure. Surgery removes as much tumour as possible: the less left, the more effective is chemotherapy and the better the prognosis. In a young woman with early disease, the uterus and other ovary may be left, for fertility; if the tumour involves both ovaries, uterus and omentum are removed.

Chemotherapy for ~ 6 months post-op is usual. Cisplatin (eg $\geq 50\text{mg/m}^2$) or carboplatin iv are used (\pm doxorubicin & cyclophosphamide; but one good trial found that these increased side-effects, not survival—eg alopecia, wcc \downarrow , nausea).² Carboplatin causes less nausea, vomiting, and neuropathy and is not renally toxic, and may not require hospital admission. Relapse is common: substituting the cyclophosphamide for paclitaxel (from Pacific yew trees) may be better. Radiotherapy may be tried.

Further treatment may involve a 'second look' laparotomy, further chemotherapy, or radiotherapy. Colloidal gold may control ascites.

Palliative care involves relief of symptoms, which are generally due to extensive peritoneal disease.

Vaginal discharge

Discharge may be physiological (eg pregnancy; sexual arousal; puberty; Pill). Most discharges are smelly, itchy, and due to infection. Foul discharge may be due to a foreign body (eg forgotten tampons, or beads in children).

Note the details of the discharge. Has she a sexually transmitted disease (STD)? See *OHCM* p 584–588. If so, refer to a genito-urinary clinic.

Do a speculum examination and take swabs: endocervical (and special medium) for chlamydia; cervical swabs for gonorrhoea (*OHCM* p 588).

► Discharges rarely resemble their classical descriptions.

Thrush (*Candida albicans*) Thrush is the commonest cause of discharge, which is classically white curds. The vulva and vagina may be red, fissured, and sore, especially if there is an allergic component. Her partner may be asymptomatic. Pregnancy, contraceptive and other steroids, immunodeficiencies, antibiotics, and diabetes are risk factors—check for glycosuria.

Diagnosis: Microscopy (shows mycelia or spores) and culture.

Treatment: A single imidazole pessary, eg clotrimazole 500mg, plus cream for the vulva (and partner) is convenient. She may need reassurance that thrush is not necessarily sexually transmitted.

Thrush is often recurrent; consider nystatin vaginal pessaries for 14 nights (messy) or fluconazole 150mg po as a single dose (avoid if pregnant). *Candida* elsewhere (eg mouth, natal cleft) of both partners may cause reinfection. Live yoghurt (on a tampon) or acetic acid jelly pH 4, eg Aci-Jel® may prevent or relieve mild attacks. Avoidance of chemicals around the vulva (no bathsalts); wiping the vulva from front to back; cotton underwear, double rinsed; and vinegar baths, may help.

Trichomonas vaginalis (tv) This produces vaginitis and a thin, bubbly, fishy smelling discharge. It is sexually transmitted. Exclude gonorrhoea, which often coexists. The motile flagellate may be seen on a wet film ($\times 400$ magnification), or cultured. **Treatment:** Metronidazole 400mg/12h po for 5 days or 2g po stat; treat the partner; if pregnant, use the 5-day regimen.

Bacterial vaginosis Prevalence $\sim 10\%$ (uk), most asymptomatic. If discharge, it has fishy odour, from cadaverine and putrescine. Vaginal pH is >4.5 . The vagina is not inflamed and pruritus is uncommon. Mixed with 10% potassium hydroxide on a slide, a whiff of ammonia may be detected. Stippled vaginal epithelial 'clue cells' may be seen on wet microscopy. There is altered bacterial flora—overgrowth, eg of *Gardnerella vaginalis*, *Mycoplasma hominis*, peptostreptococci, *Mobiluncus* and anaerobes, eg *Bacteroides* species—with too few lactobacillae. There is increased risk of preterm labour, intra-amniotic infection in pregnancy, susceptibility to HIV-1,¹ and post-termination sepsis. **Diagnosis:** By culture.

Treatment: Metronidazole 2g po once, gel pv, or clindamycin 2% vaginal cream, 1 applicatorful/night pv 7 times. If recurrent, treating the partner may help. If pregnant, use metronidazole 400mg/12h po for 5 days.

Discharge in children may reflect infection from faecal flora, associated with alkalinity from lack of vaginal oestrogen (prepubertal atrophic vaginitis). *Staphs* and *streps* may cause pus. *Threadworms* cause pruritus. Always consider sexual abuse. Gentle rectal examination may exclude a foreign body.

Tests: Do a vulval \pm vaginal swab. msu: is there glycosuria? For prolonged or bloody discharge, examination under anaesthesia may be needed (a paediatric laryngoscope can be used as a speculum) \pm ultrasound or x-rays.

Management Discuss hygiene. If an antibiotic is needed, erythromycin is a good choice. An oestrogen cream may be tried (≤ 1 cm strip).

Pelvic infection

Pelvic infection affects the Fallopian tubes (salpingitis) and may involve ovaries and parametra. 90% are sexually acquired, mostly chlamydia: 60% of these are asymptomatic (♀/♂ = 1:1) but infertility or ectopic pregnancy may be the result, *which is why screening has been proposed*—eg by a urine ligase chain reaction DNA: see opposite. Other causes, eg the gonococcus are rarer. Organisms cultured from infected tubes are commonly different from those cultured from ectocervix, and are usually multiple. 10% follow childbirth or instrumentation (insertion of IUCD, TOP) and may be streptococcal. Infection can spread from the intestinal tract during appendicitis (Gram -ve and anaerobic organisms) or be blood-borne (tuberculosis).

Salpingitis Patients with *acute salpingitis* may be most unwell, with pain, fever, spasm of lower abdominal muscles (she may be most comfortable lying on her back with legs flexed) and cervicitis with profuse, purulent, or bloody vaginal discharge. Heavy menstrual loss suggests endometritis. Nausea and vomiting suggest peritonitis. Look for suprapubic tenderness or peritonism, cervical excitation, and tenderness in the fornices. It is usually bilateral, but may be worse on one side. *Subacute infection* can be easily missed, and laparoscopy may be needed to make either diagnosis.

Management ▶ Prompt treatment and contact-tracing minimizes complications. Take endocervical and urethral swabs if practicable. Remember to check for chlamydia. Admit for blood cultures and iv antibiotics if very unwell (eg cefoxitin 2g/6h slow iv with doxycycline 100mg/12h po) initially, then doxycycline 100mg/12h po and metronidazole 400mg/12h po until 14 days treated. Seek advice from microbiologist if gonorrhoea isolated. If less unwell give ofloxacin 400mg/12h po and metronidazole 400mg/12h po for 14 days. Trace contacts and ensure they seek treatment (seek help of the genito-urinary clinic).

Complications If response to antibiotics is slow, consider laparoscopy. She may have an abscess (draining via the posterior fornix prevents perforation, peritonitis, and septicaemia—but laparotomy may be needed). Inadequate or delayed treatment leads to chronic infection and to long-term tubal blockage (8% are infertile after 1 episode, 19.5% after 2, 40% after 3). Advise that barrier contraception protects against infection. Ectopic pregnancy rate is increased 10-fold in those who do conceive.

Chronic salpingitis Unresolved, unrecognized, or inadequately treated infection may become chronic. Inflammation leads to fibrosis, so adhesions develop between pelvic organs. The tubes may be distended with pus (pyosalpinx) or fluid (hydrosalpinx).

Pelvic pain, menorrhagia, secondary dysmenorrhoea, discharge, and deep dyspareunia are some of the symptoms. She may be depressed. Look for tubal masses, tenderness, and fixed retroverted uterus. Laparoscopy differentiates infection from endometriosis.

Treatment is unsatisfactory. Consider long-term broad-spectrum antibiotics (eg tetracycline 250mg/6h po 1h before food for 3 months), short-wave diathermy and analgesia for pain, and counselling. The only cures are the menopause or surgical removal of infected tissue.

Urine screening tests to prevent chlamydial pelvic infection¹

Current proposals are for opportunistic screening in family planning and TOP contexts, and routine GP appointments for all sexually active women aged up to 25yrs. This might miss up to 20% of infections (increasing the age limit to 30 might miss just 7%).¹ It is not clear how this screening is to be woven into routine appointments (which are already overfull), and whether patients will find screening acceptable.

1 F Boag 1998 *BMJ* i 1474

Endometriosis

Essence Foci of endometrial glandular tissue, looking like the head of a burnt match, are found beyond the uterine cavity, eg on an ovary ('chocolate cyst'), in the rectovaginal pouch, uterosacral ligaments, on the surface of pelvic peritoneum, and rarely in the umbilicus, lower abdominal scars, and distant organs, eg lungs. If foci are found in the muscle of the uterine wall, the term *adenomyosis* is used. It occurs in ~12% of women; not all have symptoms, and in the asymptomatic, it may be physiological.

Cause Possibly cell rests, or retrograde menstruation (Sampson's theory, explaining its association with age—typically 40–44yrs, long duration of iucd and tampon use, and, possibly, tight underwear;¹ its negative association with pregnancy and the Pill, and its distribution in the pelvis—but not its appearance elsewhere). There is a genetic component²—and there is an autoantibody association. Endometriotic foci are under hormonal influence with waning in pregnancy and at the menopause—and bleeding during menstruation. Free blood irritates, provoking fibrosis and adhesions.

Presentation Endometriosis may be asymptomatic, even though extensive. Pelvic pain is the commonest symptom (classically cyclical, at the time of periods). It may be constant, eg if due to adhesions. Secondary dysmenorrhoea and deep dyspareunia are common. Periods are often heavy and frequent, especially with adenomyosis. Patients may present with infertility. Extra-pelvic endometriosis presents with pain or bleeding at the time of menstruation at the site of the pathology, eg haemothorax.

Diagnosis *Per vaginam*: fixed retroverted uterus or nodules in the uterosacral ligaments and general tenderness suggest endometriosis. An enlarged, boggy, tender uterus is typical of adenomyosis. MRI may help. Typical cysts or peritoneal deposits at laparoscopy differentiates it from chronic infection.

Treatment (see p 1) If asymptomatic, do not treat. Otherwise, try surgery or hormones (if not pregnant or lactating). In the UK, consider suggesting joining the Endometriosis Society,³ as treatment can be long and difficult.

Hormonal treatment aims to suppress ovulation for 6–12 months during which some (non-GI) lesions atrophy. **Danazol** up to 400–800mg/24h po is effective but SE may be unacceptable at high doses (it is a testosterone derivative with anabolic and androgenic effects so expect weight gain, acne, greasy skin, hirsutism, bloating, and fluid retention). Advise barrier contraception as it masculinizes the female fetus. **Gestrinone** 2.5–5mg po twice weekly is an alternative. LHRH analogues, eg **buserelin** nasal spray 300µg/8h or **goserelin** 3.6mg by monthly sc injection produce a reversible artificial menopause but bone demineralization occurs. SE: menopausal symptoms, headaches, depression, libido↓, breast tenderness, drowsiness, acne, dry skin, ovarian cysts (if so, stop), urticaria, changes in body hair, osteoporosis—so only give for <6 months. An alternative gonadotrophic-releasing hormone agonist with similar side-effects is **leuprorelin acetate**—dose: 3.75mg every 4 weeks IM or sc starting during the 1st 5 days of a cycle, for ≤6 months. Endometriotic resolution is similar to danazol, but SEs are less (flushes, headaches, vaginal dryness, emotional lability). Progestogens, eg **norethisterone** (Primolut N®) 5–10mg/12h po continuously for 9 months may suppress symptoms by mimicking pregnancy but is rarely very effective. *Surgical treatment* ranges from local excision or bipolar diathermy/laser of endometriotic tissue, to total hysterectomy + bilateral salpingo-oophorectomy, depending on lesion site and wishes for future fertility. Surgery is probably the best option if symptoms seriously impinge on a patient's life.

Prognosis Endometriosis is chronic or relapsing, being progressive in 50%. Surgery only minimally helps fertility (and only if mild disease).

Prolapse

A prolapse occurs when weakness of the supporting structures allows the pelvic organs to sag within the vagina. The weakness may be congenital, but it usually results from stretching during childbirth. Poor perineal repair reduces support (p 146). Weakness is exacerbated by menopausal atrophy and by coughing and straining. They may cause distressing incontinence and be a nuisance but are not a danger to health—except for third degree uterine prolapse with cystocele when ureteric obstruction can occur.

Types of prolapse are named by the structures sagging. Several types may coexist in the same patient.

Cystocele The upper front wall of the vagina, and the bladder which is attached to it, bulge. Residual urine within the cystocele may cause frequency and dysuria.

Urethrocele If the lower anterior vaginal wall bulges, this will displace the urethra and impair the sphincter mechanisms (p 70), so leading to stress incontinence. Does she leak when she laughs?

Rectocele The middle posterior wall, which is attached to rectum, may bulge through weak levator ani. It is often symptomless, but she may have to reduce herniation prior to defecation by putting a finger in the vagina.

54 Enterocoele Bulges of the upper posterior vaginal wall may contain loops of intestine from the pouch of Douglas.

Uterine prolapse With *first degree prolapse* the cervix stays in the vagina. In *second degree prolapse* it protrudes from the introitus when standing or straining. With *third degree prolapse* (procidentia) the uterus lies outside the vagina. The vagina becomes keratinized and the cervix may ulcerate.

Symptoms: 'Dragging' or 'something coming down' is worse by day. Cystitis, frequency, stress incontinence, and difficulty in defecation may occur depending on the type of prolapse. Examine vaginal walls in left lateral position with a Sims' speculum, and ask the patient to bear down to demonstrate the prolapse. Do urodynamic studies (p 71) to exclude detrusor instability and to assess voiding.

Prevention: Lower parity; better obstetric practices, pelvic floor exercises.

Treatment: Mild disease may improve with reduction in intra-abdominal pressure, so encourage her to lose weight, stop smoking, and stop straining. Improve muscle tone with exercises or physiotherapy, and, if postmenopausal, topical oestrogens, eg estriol cream 0.1% as often as required (try twice weekly).

Severe symptomatic prolapse is best treated surgically. Incontinence needs to have the cause treated (so arrange urodynamic studies to plan the best type of surgery). Repair operations (p 72) excise redundant tissue and strengthen supports, but reduce vaginal width. Is she sexually active? If so, surgery must compromise between reducing prolapse and maintaining width. Marked uterine prolapse is best treated by hysterectomy.

Ring pessaries may be tried as a temporary measure or for the very frail. Select size by diameter of vagina at level of the fornices; it will only stay in if the vagina narrows nearer the introitus. Insert into the posterior fornix and tuck above the pubic bone (easier if the ring has been softened in hot water first). Problems: discomfort, infection, ulceration (change 6–12 monthly). For those in whom they keep falling out, try a shelf pessary.

Infertility: causes and tests

► *Infertility can be devastating to both partners: its investigation a tremendous strain. Sympathetic management is crucial. 90% of young couples having regular intercourse conceive within a year. Fertility decreases with age. In the 10% of subfertile couples consider:*

- Is she producing ova? (Anovulation causes 21%.)
- Is he producing enough, healthy sperm? (Male factors cause 24%.)
- Are ova and sperm meeting? (Tubal cause 14%, hostile mucus 3%, sexual dysfunction 6%.) Aim for intercourse 3 times/week throughout the cycle.
- Is the embryo implanting?

Endometriosis is thought to be the cause in 6%. The cause is 'unexplained' in 27% of couples. With 'unexplained' infertility 60–70% of women will achieve conception within 3 years.

Initial management It takes two to be infertile. See both partners. Advise both to stop smoking and for the lady to take 0.4mg folic acid daily whilst trying to conceive. *Ask her about:* Menstrual history, previous pregnancies and contraception, history of pelvic infections or abdominal surgery, drugs. Beware retained bones after terminations in West Africans (see on ultrasound; removal usually cures).¹ *Ask him about:* Puberty, previous fatherhood, previous surgery (hernias, orchidopexy, bladder neck surgery), illnesses (venereal disease and adult mumps), drugs, alcohol intake, job (is he home when ovulation occurs?). *Ask both about:* Technique, frequency, and timing of intercourse (nonconsummation is a rare problem); feelings about infertility and parenthood; previous tests.

Examination Check the woman's general health and sexual development and examine the abdomen and pelvis. If the sperm count is abnormal, examine the man for endocrine abnormalities, penile abnormalities, varicoceles; confirm there are 2 normal testes.

Tests for ovulation If cycles are regular ovulation is likely. Basal body temperature rises midcycle (but charting is difficult and may raise anxieties).

Blood tests Check rubella status; immunize if nonimmune. If you suspect anovulation check: ● Serum progesterone 7 days before expected period (>30nmol/L is indicative of ovulation) ● Blood prolactin (if high may be due to prolactinoma; MRI of brain and pituitary gland) ● FSH (>10u/L indicates a poor response to ovarian stimulation; it may indicate primary ovarian failure—but FSH is pulsatile in release and one may get a high reading depending on the stage of the cycle) ● LH (for polycystic ovary syndrome) ● TFT.

Semen analysis Unless there has been a satisfactory postcoital test, analyse semen after 3 days' abstinence for: ● Volume (mean 2.75mL now, formerly 3.4mL in the 1940s) ● Sperm count/morphology. ● Antibodies. ● Infection. Normal count >20 million sperm/mL, >50% motile and >60% normal form; WBC < 1 million/mL; mean sperm count = 66 million/mL (113 million/mL in the 1940s; this average is falling—which may be due to an environmental influence—or more frequent ejaculations. Examine 2 specimens as variation may be considerable. Transport sample fresh to lab (masturbated into a wide topped container). Avoid temperatures <15°C or >38°C. Reduced counts require specialist referral.

Subfertility options: abbreviations, problems, and ethics¹

DI	Donor insemination (in UK law, children have no rights to information about their father: this can lead to problems—eg most children want their fathers to be proud of them, and with DI this is impossible.) ² The great metaphysical questions such as <i>where do I come from</i> become more opaque than ever.
FEC	Fetal egg child (offspring from an egg taken from the ovary of an aborted fetus).
ICSI	Intracytoplasmic sperm injection (directly into an egg). If necessary, the sperm may be obtained surgically from the testis or epididymis. Such patients, with no exposure to their partner's sperm, are at increased risk of pre-eclampsia (p 96): this supports a model of pre-eclampsia postulating absence of partner-specific immune tolerance as a causative factor. ³
IUIS	Intrauterine insemination with superovulation: a USA favourite. In 20%, this causes twins (in 10%, higher order pregnancies when pregnancy is achieved; a possibly unacceptable disadvantage). ⁴
IVF	<i>In vitro</i> fertilization
GIFT	Gamete intrafallopian transfer
MESA	Microepididymal sperm aspiration from testis, post-vasectomy
OT/NT(P)	Ooplasmic transfer/nuclear transfer procedure: the baby has 2 mothers: one (too old to conceive normally) provides a nucleus; the other provides fresher cytoplasm (+ mitochondrial DNA) for the ovum. This is an example of human germline modification ⁶ which is legal in the USA, but illegal in the UK. ⁵
PESA	Percutaneous epididymal sperm aspiration (rather like MESA, but using a 22G butterfly needle inserted into the epididymis—so scrotal exploration is not required).
PHC	Pregnancy by human cloning (illegal in many countries partly because of concern about premature ageing—but achieved by Italian doctors, up to 8 weeks gestation, in April 2002)
POST	Peritoneal oocyte sperm transfer
POT	Pregnancy by ovary transplantation (China leads the field in ovarian transplantation)
SUZI	Subzonal sperm injection (directly into an egg)
TET	Tubal embryo transfer
TUFT	Transuterine fallopian transfer
ZIFT	Zygote intrafallopian transfer

Ethical issues

Various national embryology authorities exist and pronounce on the ethical dimensions of fertility treatments—and their edicts can appear to be set in stone (although being mutually contradictory with those from other countries); one problem with this approach is that fertility treatments are constantly changing, as is society's views on what is acceptable—and it is not clear whether these views should lead, or simply be taken into account (an opaque phrase) or be trumped by appeal to some higher authority (God, or the conscience of a quango).

Infertility: tests and treatment

Tests of tubal patency (Screen for chlamydia first).

- 1 Laparoscopy with dye. Pelvic organs are visualized and methylene blue dye injected through the cervix: if tubes are blocked proximally they do not fill with dye; with distal block there is no 'spill' into the peritoneal cavity.
- 2 A hysterosalpingogram (contrast x-ray) demonstrates uterine anatomy and tubal 'fill and spill'. Unpleasant, it may require premedication. False positives may occur with tubal spasm. Give antibiotics, eg cefradine 500mg/8h po with metronidazole 1g/12h pr for 24h before and eg 400mg/12h po for 5 days after procedure, to prevent pelvic infection.

The postcoital test Taken in the immediate pre-ovulatory phase, 6–12h after intercourse, cervical mucus is removed and examined under high-power ($\times 400$) microscopy. A satisfactory test (ovulatory mucus, >10 motile sperm/high-power field) shows that his sperm are adequate, coitus is effective and the cervical mucus does not contain antibodies. Typical use: if sexual dysfunction suspected or no semen sample provided by the male.

Treatment of infertility Treatment is directed at the cause.

Azoospermia is unresponsive to treatment. A low sperm count may be improved by avoiding tobacco and alcohol. Avoiding tight pants was advised, but experiments show no change in testis T° or semen parameters. Will they consider donor insemination (AID = artificial insemination by donor)?

Problems of sperm deposition (eg impotence) can be circumvented by artificial insemination using the partner's sperm.

Hyperprolactinaemia (OHCM p 312) Remove the cause if one is found (pituitary macro-adenoma, drugs); if not, give bromocriptine 2.5mg/24h po, increasing slowly until blood prolactin is normal. (Side effect: nausea.)

Anovulation is managed by stimulating follicle development using clomifene 50–150mg/24h po on days 2–6 inclusive. SE: flushes (10%), visual disturbance (1.5%), abdominal pain (5.5%—see p 75); warn about risk of multiple pregnancy \pm excess risk of ovarian cancer (p 42). Monitor the response to clomifene with luteal phase progesterone. Human chorionic gonadotrophin (HCG) resembles LH and may be needed to rupture a ripe follicle. If clomifene does not help, gonadotrophin injections or LHRH analogues may be tried.

Tubal problems may be remedied by surgery but results are poor.

Assisted fertilization Psychological stability (and finance) are required. The 'take-home baby rate' averages 10% and fetal loss, ectopic pregnancy, obstetric problems, multiple births, and fetal abnormality are common.

In vitro fertilization (IVF) is used for tubal (and other) problems. The ovaries are stimulated (see *hyperstimulation* p 75), and ova are collected (by transvaginal aspiration under transvaginal ultrasound guidance), fertilized, and embryos are returned to the uterus as an outpatient procedure.

Gamete intrafallopian transfer (GIFT) may be tried where tubes are patent, eg 'unexplained infertility'. Gametes are placed in the tubes by laparoscopic cannulation. Pregnancy rate $\sim 28\%$ per cycle treatment. ZIFT (zygote or early cleaved embryo transfer) can give higher pregnancy rates of up to 48% in women with unexplained infertility.

If there is severe oligospermia *intracytoplasmic sperm injection* may be possible (a spermatozoa is injected directly into the oocyte's cytoplasm). The source of sperm is the epididymis or testis in men with obstructive azoospermia; even if the problem is non-obstructive, sperm can be retrieved in $\sim 50\%$.

The possibility of adoption should not be forgotten. Those who remain childless may value counselling or a self-help group.

Male infertility

Spermatogenesis takes place in the seminiferous tubules. Undifferentiated diploid germ cells (spermatogonia) multiply and are then transformed into haploid spermatozoa. The total duration of a cohort of spermatogonia to develop into spermatozoa is 74 days. FSH and LH are both important for initiation of spermatogenesis at puberty. LH stimulates Leydig cells to produce testosterone. Testosterone and FSH stimulate Sertoli cells to produce essential substances for metabolic support of germ cells and spermatogenesis.

Spermatozoa A spermatozoon has a dense oval head (containing the haploid chromosome complement), capped by an acrosome granule (contains enzymes essential for fertilization) and is propelled by the motile tail. Seminal fluid forms 90% of ejaculate volume and is alkaline to buffer vaginal acidity. Only about 200 sperm from any ejaculate reach the middle third of the Fallopian tube, the site of fertilization.

Male infertility Male factors are the cause of infertility in ~24% of infertile couples. In reality, most are subfertile. Only a small number of men have an identifiable cause. Causes include (% cited in one study):

- **Idiopathic oligo/azoospermia (16%):** Testes are usually small and FSH ↑.
- **Asthenozoospermia/teratozoospermia (17%):** In asthenozoospermia sperm motility is reduced due to structural problems with the tails. Teratozoospermia indicates an excess of abnormal forms.
- **Varicocele (17%):** this is controversial for varicocele is found in 15% of males, most of whom have normal fertility.
- **Genital tract infection (4%):** gonococci, chlamydia, & Gram -ve enterococci cause adnexal infection (± painful ejaculation, urethral discharge, haemospermia, dysuria, tender epididymes, tender boggy prostate). Confirm by semen culture, urethral swab or finding $>10^6$ peroxidase +ve polymorphs/mL semen. Treatment has not been shown to restore fertility.
- **Sperm autoimmunity (1.6%):** Risk factors for antibodies: vasectomy, testis injury, genital tract obstruction, family history of autoimmunity. Most are on sperm membranes or in seminal fluid, but may occur in the woman.
- **Congenital (cryptorchidism, chromosome disorders—2%):** Klinefelter's account for 50% chromosome disorders. For optimal fertility undescended testes should be fixed in the scrotum before 2yrs of age.
- **Genital tract obstruction (1.8%):** Azoospermia, normal sized testes with normal or high FSH suggests this. It may follow infection, vasectomy, or be congenital (as in cystic fibrosis). It may be amenable to surgery, eg epididymovasostomy to bypass epididymal obstruction.
- **Systemic—eg iatrogenic, or from drugs, eg cannabis (1.3%).** medline:6090909
- **Coital disorders (1%).**
- **Gonadotrophin deficiency (0.6%):** this is the only cause of testicular failure consistently treatable by hormone replacement.

Examination Look at body form and secondary sexual characteristics. Any gynaecomastia? Normal testicular volume is 15–35mL (compare with Prader orchidometer[□]). Rectal examination may reveal prostatitis.

Tests Semen analysis (p 56). Postcoital tests with midcycle cervical mucus may indicate antibody presence. Plasma FSH distinguishes primary from secondary testicular failure. Testosterone and LH levels are indicated if you suspect androgen deficiency. Agglutination tests to detect antibodies are available.

Treatment Intracytoplasmic sperm injection (direct into egg) is the main tool for most male infertility.

Contraception

► Any method, even *coitus interruptus*, is better than none. 90% of young women having regular unprotected intercourse become pregnant within a year. Properly used, contraception reduces this to <1% (combined Pill, injectables, sterilization); around 2% (POP, IUCD, cap, sheath); 12% (mucothermic)—success rates may be much higher; 15% (withdrawal). Failure rates may be much higher for user-dependent methods.

Barrier methods ► The main reason for failure is not using them.

Sheaths reduce transmission of most STDs but not those affecting the perineum. Caps give some protection against gonorrhoea and chlamydia but not syphilis or herpes. Some spermicides inactivate HIV *in vitro*.

- **Sheaths** are effective when properly used and with spermicides. Unroll onto the erect penis with the teat or end (if teatless) pinched to expel air. This prevents bursting at ejaculation.
- **Caps** come in several forms. Diaphragms stretch from pubic bone to posterior fornix. She must check after insertion that the cervix is covered. Cervical caps fit over the cervix (so need a prominent cervix). Insert <2h before intercourse (keep in place >6h after sex). Use with a spermicide. Some find them unaesthetic. Problems: UTIs, rubber sensitivity.
- **Cervical sponges** These are simple but often fail.
- **The female condom** (eg Femidom®) Prescription and fitting are not needed. It has not proved popular. One reason for failure is that the penis goes alongside it, rather than in it; another, that it gets pushed up in the vagina or may fall out. They can be noisy.
- **Spermicides**—Unreliable unless used with a barrier.

Natural (mucothermal) methods involve monitoring physiological phenomena to find fertile times (6 days prior to ovulation; the life of a sperm) to 2 days afterwards (the life of the ovum). Cervical mucus becomes clear and sticky at the beginning of the fertile time and dry at ovulation (but is altered by semen and vaginal infections). Basal body temperature $\uparrow \sim 0.3^\circ\text{C}$ after ovulation (affected by fevers, drugs, recent food, or drink). Additional observations (mittelschmerz, $p\ 74 \pm$ cervix changes) improve accuracy. Success is common if: • Regular cycles • Dedication • Self-control. UK teachers are available (tel. 01222 754628).

High-technology natural methods Devices such as Persona® use solid-state urine chemistry on sticks for dual measurements of the oestrogen (o)estrone-3-glucuronide (E3G—peaks 24h pre-ovulation) and luteinizing hormone (LH—ovulation occurs within 36h of LH surge and sperm penetration through cervical mucus drops after the surge). Microtechnology builds up a database of the woman's natural variability over time, and this is used to give her either a green light (almost infertile), a red light (fertile—typically days 6–10) or an orange light (do an early-morning urine test for E3G and LH). Usually, only 8 urine tests are needed per cycle. She needs to purchase sticks and one monitor (this houses a button to press the morning her period starts: she needs to check the lights on the monitor before passing urine each morning, in case a test is needed). **Reliability:** 93–95% (manufacturer's data, in motivated patients: it may be much less in practice; results must be regarded as only preliminary: explain uncertainty to your patient). **CI:** Cycle <23 or >35 days or variation >10 days; breast feeding; if already on hormones or tetracycline (minocycline is OK); menopausal; liver or kidney disease; polycystic ovary disease, or if pregnancy is definitely unwanted.

The ideal contraceptive—and the realities

An ideal contraceptive is: 100% effective, with only *desirable* side-effects (eg protection from sexually transmitted disease)—and it must be *readily reversible*, and be usable *unsupervised by professionals*. Try to find the best compromise for each person depending on age, health, and beliefs. Methods available:

- 'Natural methods' (no intercourse near time of ovulation)—acceptable to Catholic Church; also, the simplest are free, requiring no 'pollution of the body' with drugs—see opposite.
- Barrier methods (low health risk but need high user motivation—with some protection to HIV). See opposite.
- Hormonal (complex health risks but highly effective, p 64–8).
- IUCD (convenient and effective—if not contraindicated—p 62).
- Sterilization (very effective but effectively irreversible, p 69).

'Is she pregnant already?'

This is a frequent question in family planning and other clinics. If a pregnancy test is not available, women who may be pregnant already will often be denied the contraception they need. Here, consider using this check list to see if the patient may be pregnant. If she answers Yes to **any** of these questions, and she is free from signs or symptoms of pregnancy, then pregnancy is very unlikely (negative predictive value >99%).

- Have you given birth in the past 4 weeks?
- Are you <6 months postpartum **and** fully breastfeeding, **and** free from menstrual bleeding since you had your child?
- Did your last menstrual period start within the last 7 days?
- Have you been using a reliable contraceptive consistently and correctly?
- Have you not had sex since your last period?

medline:10470704

Real contraceptives—the failure rates

No method used	8500	pregnancies per 10 000 women using/year
Cervical cap	3000	
Spermicides	2100	
Female condom	2100	
Withdrawal	2000	
Male condom	1200	
POP (progesterone-only Pill)	300	
Copper coil	40	
Injectable contraceptive	30	
Tubal ligation	17	
Combined Pill	8	
Vasectomy	4	

The intrauterine contraceptive device (IUCD)

IUCDs (loops, coils) are plastic shapes ~3cm long around which copper wire is wound, and carrying a plastic thread from the tail, eg Novagard®. They inhibit implantation and may impair sperm migration. Most need changing every 3–5 years. They have replaced the larger, noncopper-bearing 'inert' types (eg Lippes loop®), which caused more complications but did not need changing (and so are sometimes still found *in situ*).

Most of those who choose the IUCD (6%) are older, parous women in stable relationships, in whom the problem rate is low.

Problems with IUCDs **1** They tend to be expelled by a uterus which is nulliparous or distorted (eg by fibroids). **2** Ectopic pregnancy is more likely. **3** They are associated with pelvic infection and infertility, following sexually transmitted disease—or sometimes introduced during insertion. **4** They tend to produce heavy, painful periods. **Contraindications:** Aim to exclude women at high risk of these problems, also those who are immunosuppressed (eg HIV+ve), have Wilson's disease (OHCM p 224), or those with cardiac lesions (unless the coil is inserted under antibiotic cover).

Insertion Skilled insertion minimizes complications. Each device has its own technique, so read the instructions carefully and practise beforehand.

An IUCD can be inserted any time (and as postcoital contraception), as long as she's not pregnant, but it is more likely to be expelled during a period or shortly after birth. Determine the position of the uterus. Then insert a uterine sound to assess the cavity length. Then insert the IUCD, placing it in the fundus. This may cause cramps. Once the coil is in place, the threads are cut to leave 3cm visible in the vagina. Teach her to feel the threads after each period. ► Insertion of IUCDs may provoke 'cervical shock' (from increased vagal tone). Have IV atropine available.

Follow-up The first 3 months are the peak time for expulsion and the coil should be checked after the next period, then 6-monthly. Threads are often easier to feel than to see.

Lost threads The IUCD may have been expelled, so advise extra contraception and exclude pregnancy. Then sound the uterus. If you find the coil you may be able to bring down the threads with a special instrument. If you cannot find the threads, they may have dropped off. If you cannot find the coil, seek it on ultrasound. A coil may perforate the uterus, usually after insertion and often with little pain (remove, eg laparoscopically).

Infection This can be treated with the device in place, but if removed do not replace it for 6 months. With symptomatic Actinomyces, remove coil, cut off threads and send it for culture. If positive, seek expert advice on treatment.

Pregnancy >90% are intrauterine. Remove coil, if you can as soon as pregnancy is diagnosed to reduce risk of miscarriage (20% if removed early, 50% if left), and to prevent risk of late septic abortion.

Removal Alternative contraception should be started (if desired) prior to removal, as otherwise there may be a fertilized ovum in the tubes. At the menopause, remove after at least 6 months' amenorrhoea.

IUCDs carrying hormones—eg Mirena® (carries levonorgestrel). Local effects make implantation less likely, and periods lighter; perhaps 20% may experience reversible amenorrhoea (reliability matches that of sterilization, see p 69). It lasts ~5 years. There may be less risk of ectopic pregnancy. Risk of sexually transmitted infection is reduced. Warn about spotting ± heavy bleeding (NB: bleeding may become scanty or absent after a few cycles).

Emergency contraception

This is for use after isolated episodes of unprotected intercourse, eg 'the split sheath' and should not be used regularly.

Management History of LMP; normal cycle; number of hours since unprotected intercourse; history of personal thromboembolism or focal migraine. Check BP. Explain that teratogenicity has not been demonstrated. Discuss future contraception. Give supply of oral contraceptives if day 1 start at next period is planned. Arrange follow-up at 3–4 weeks.

Levonorgestrel This is with progesterone-only pills initiated within 72 hours of unprotected intercourse. It is suitable for those with focal migraine and past thromboembolism. It is not suitable for those with acute active porphyria. Nausea occurs in 16%, vomiting in 2%. Two doses of 0.75mg levonorgestrel ie one tablet of Levonelle-2[®], (equivalent to the unlicensed use of 25 Microval[®] or Norgeston[®] tablets or 20 tablets Neogest[®], no longer endorsed by the Family Planning Association) taken 12h apart. The failure rate is about 1.1%. The earlier it is taken after unprotected intercourse, the fewer the pregnancies which occur. Those on liver enzyme inducers eg rifampicin, rifabutin, griseofulvin, phenytoin, tropicamide, barbiturates including primidone, ritonavir, double the first dose taken; but keep the second dose the same.

When prescribing post-coital hormonal contraception warn that the first period may be early or late; that barrier contraception should be used until the next period; and that she should return if she suffers any lower abdominal pain or the next period is abnormal.

Recent UK initiatives: Levonelle-2[®] may now be bought 'over the counter'—and lessons on its use are being conducted in schools, particularly emphasizing the 72h time limit, rather than just 'the morning after'.¹

The emergency IUCD More effective than hormonal contraception, a copper IUCD can be inserted within 120 hours of unprotected intercourse. If exposure was more than 5 days previously it can be inserted up to 5 days after likely ovulation, so is useful in women who present later. Failure rates are extremely low. Infection is a risk (eg after sexual assault) and should be screened for, if suspected. It is possible to insert under antibiotic cover. It inhibits implantation.

Hormonal contraception (the Pill)

3 million women in the UK take the combined oral contraceptive (CoC). CoCs contain oestrogen (eg ethinylestradiol) with a progestogen, either in fixed ratio or varying through the month (phased). Low-dose Pills ($\leq 30\mu\text{g}$ oestrogen) are the norm. The combined Pill is taken daily for 3 weeks followed by a week's break. This inhibits ovulation and gives a withdrawal bleed in the Pill-free week. When prescribing CoCs, pay attention to these areas:

History Why does she want the Pill? Does she know about risks? These must be explained. Has she considered the alternatives? Are there any contraindications in her or her family, eg thrombotic disorders? Ask about smoking (see below) and drugs, and about contact lenses (Pill use may cause dry eyes). Does she have any anxieties, eg weight-gain? Can she be helped to stop smoking?

Contraindications These are legion; check British National Formulary (BNF). Obesity (body mass index BMI $>39\text{kg/m}^2$); diabetes with complications; BP $>160/100$; smoking $>40/\text{day}$; age $>50\text{yrs}$; migraine as p 65; past history of venous or arterial thrombosis; thrombophilia p 151; heart disease with pulmonary hypertension or embolus risk; transient ischaemic attacks; liver disorders (adenoma, gallstones, Dubin-Johnson and Rotor syndromes, pre-recovered hepatitis); porphyria; systemic lupus erythematosus; past haemolytic uraemic syndrome, pruritus or cholestatic jaundice of pregnancy, pemphigoid gestationis, hydatidiform mole (until experts advise safe); chorea; deteriorating otosclerosis; breast or genital tract tumours; breastfeeding ($<6\text{months}$, not weaning off), undiagnosed vaginal bleeding; immobility (in bed/leg in plaster cast).

Cautions Consult BNF. Hyperprolactinaemia (unless specialist advice); sickle cell disease; inflammatory bowel disease (Crohn's too); severe depression. *Venous risk factors* Prescribe with caution if one of the following; avoid if 2 or more: first degree relative family history of thromboembolism (avoid gestodene and norgestrel containing pills); obesity (BMI $>30\text{kg/m}^2$); varicose veins (avoid during sclerotherapy); immobility (wheel-chair). *Arterial risk factors* Prescribe with caution if one of the following; avoid if 2 or more: smoker; age $>35\text{yrs}$; migraine without aura; obesity (BMI $>30\text{kg/m}^2$); diabetes mellitus; BP $>140/90$; family history of first degree relative arterial disease $<45\text{yrs}$ (avoid if known atherogenic lipid profile).

Drugs interfering with the Pill *Liver enzyme inducers*, eg anticonvulsants, griseofulvin, and rifampicin reduce efficacy by \downarrow circulating oestrogen. If short-term use, take extra precautions (p 66) whilst used and for 7 days after (4 weeks for rifampicin). With long-term use consider using 50 μg oestrogen Pills with higher doses of progesterone, eg Norinyl-1[®]. Use another type of contraceptive if on long-term rifampicin. *Antibiotics*: p 66.


Benefits of the Pill Very effective contraception; lighter, less painful 'periods'; reduced premenstrual syndrome. In long-term users, reduced risk of ovarian tumours (carcinoma $\downarrow 40\%$) and endometrial carcinoma ($\downarrow 50\%$); less pelvic infection and endometriosis.

Serious disadvantages Risks of arterial and venous disease are increased, eg DVT and myocardial infarction. Risk of death from the Pill increases sharply in those $>40\text{yrs}$: 1:2500 for nonsmokers; 1:500 for smokers. *Breast cancer*: In one large retrospective study ($n=9257$) in women aged 35–64yrs, current or former Pill use was not associated with a significantly increased risk of breast cancer. But in younger women, rates may rise from $\sim 0.2\%$ to $\sim 0.3\%$.

Migraine, ischaemic stroke, and the Pill¹

The problem is ischaemic stroke. The background annual incidence is 2 per 100,000 women aged 20, and 20 per 100,000 for those aged 40. Migraine itself is a risk factor. For those with migraine and cōc use, incidence of ischaemic stroke becomes 8:100,000 if aged 20; and 80:100,000 in those aged 40. Low-dose cōcs only should be used. Those with migraine with aura are known to be at special risk precluding use of combined Pills in these women (however there is no problem with them using progestosterone only or nonhormonal contraception). Other risk factors for ischaemic stroke include smoking, age >35yrs, raised BP, obesity (body mass index >30), diabetes mellitus, hyperlipidaemia and family history of arterial disease <45yrs. Women known to have migraine should be warned to stop Pills immediately if they develop aura or worsening of migraine. If a woman has 1st migraine attack on cōc, stop it, observe closely: restart cautiously only if there are no sequelae and if migraine attack was without aura² and there are no other risk factors (above).

Diagnosing migraine with aura Formerly called *classical/focal migraine*

- Slow evolution of symptoms (see below) over several minutes
- Duration of aura usually 10–30min, resolving within 1h, and typically before onset of headache
- Visual symptoms (99% of auras), eg:
 - Bilateral homonymous hemianopia
 - Teichopsia and fortification spectra, eg a gradually enlarging C 
 - Positive (bright) scotomata
- Sensory disturbance (31% of auras)
 - Usually associated with visual symptoms
 - Usually in one arm spreading from fingers to face (leg rarely affected)
- Speech disturbance (18% of auras):
 - Dysphasia; dysarthria; paraphasia
- Motor disturbances (6% of auras)

Both motor and speech disturbances are usually accompanied by visual and/or sensory disturbances.

Migraine without aura: (Formerly called *simple* or *common* migraine) includes symptoms of blurred vision, photophobia, phonophobia, generalized flashing lights affecting the whole visual field in both eyes, associated with headache.

Absolute contraindications to cōc use

- Migraine with aura
- Migraine without aura in women with >1 risk factor for stroke
- Severe migraine or status migrainosus (>72h)
- Migraine treated with ergot derivatives

¹ E McGregor 1998 *Br J Fam Plan* 24 55 ² E McGregor 1995 *Br J Fam Plan* 21 16

Hormonal contraception: further details

Do BP 6-monthly. Check weight and breasts—eg if >35yrs. Do a cervical smear if due (p 32). Adverse effects are dose-related: aim is to give the lowest dose Pill that gives good cycle control (ie no breakthrough bleeding).

Progesterone issues Norethisterone and levonorgestrel-containing COCs may cause breakthrough bleeding, acne, headaches, weight ↑, so prompting a search for newer progesterones, eg gestodene or desogestrel (Femodene®, Marvelon®, Minulet®, Mercilon®, Triadene®, Triminulet®)—but see problems with thrombosis (p 67). These may have a rôle in those without risk factors for venous thrombosis (p 64), in those with arterial risk factors (p 64), or bad acne. Overall, risk of death with the newer Pills from PE is higher than expected ($\geq 1/100,000/\text{yr}$, eg in New Zealand).

Drospirenone (DRSP; Yasmin®) is a spironolactone-like progesterone with antiandrogenic/antimineralocorticoid action (said not to ↑ weight, and to help PMS, p 16—unproven); clotting may still be a problem. medline:11559453

Switching from PoP: If to a COC, start on day 1 of period. Use condoms for 7 days. If to a progesterone-only Pill (PoP), start as the old pack finishes.

Switching from COC: If to a COC, start immediately with no Pill-free interval. If to a PoP, start immediately the COC packet is finished.

Starting COCs and PoPs Day 1 of cycle, or on the day of TOP, or 3 weeks postpartum or 2 weeks after full mobilization after major surgery. With day 1–3 COC starting regimen, contraceptive cover is immediate; use other precautions for 7 days with PoP and if COC started day 4 or 5 of period.

Stopping the Pill Tell to stop at once if she develops: •Sudden severe chest pain •Sudden breathlessness (or cough with bloody sputum) •Severe calf pain •Severe stomach pain •Unusual severe prolonged headache. See cautions on p 64; low-risk nonsmokers may continue into their 40s. On stopping, 66% menstruate by 6 weeks, 98% by 6 months; women who are amenorrhoeic after the Pill were usually so before.

Special cases If >12h late (or diarrhoea), continue Pills but use condoms too for 7 days (+ days of diarrhoea); if this includes Pill-free days, start next pack with *no break*. Use condoms during and for 7 days after antibiotics. If a tablet from the 1st 7 days in pack is forgotten, consider postcoital contraception. *Vomiting < 3h post Pill:* Take another. *Postcoital options:* p 63.

Progesterone only Pill (PoP, 'mini-pill') Low-dose progestogen renders cervical mucus hostile to sperm. In some women it also inhibits ovulation, improving efficacy but causing erratic bleeding, its worst side-effect. The risk of ectopics is increased. It is less effective than the combined Pill except in older, less fertile women. It can be used by most women with medical problems contraindicating the combined Pill, and by breast feeding mothers. Take at same time daily $\pm 3\text{h}$, otherwise extra precautions are needed for 7 days. A regular lifestyle or using an alarm watch helps.

Postnatal Start 21 days after birth: eg COC if not breast feeding; PoP, Depo-Provera® (or ?Implanon®) if breast feeding. IUCD: fit ~4 weeks postpartum.

At the menopause Stop PoP if >50yrs old with >1 year's amenorrhoea (2yrs if <50yrs old)—as a rough guide. In this last year, a spermicide and sponge is probably adequate due to declining fertility. As COC masks the menopause, aim to stop all women at 50yrs, and replace with a nonhormonal method.

Buying the Pill over the counter This might be safe for non-smokers with no hypertension who are having regular cervical smears. But it is hard to see how counter sales could be limited to this very low-risk group.

Terminology

1st generation Pills are the original Pills containing 50µg (o)estrogen. *2nd generation Pills* are those containing ≤ 35µg oestrogen and levonorgestrel, norethisterone, norgestimate, or cyproterone acetate. *3rd generation Pills* contain desogestrel, or gestodene as the progesterone. Although designed to be more lipid friendly 3rd generation Pills have not been proven to be better in those with cardiac risk factors and are more thrombogenic.¹

Risk of thrombosis¹

Not on Pill	5:100,000
2 nd generation Pill	15:100,000
3 rd generation Pill	25:100,000
Pregnancy	60:100,000

Carriage of factor V Leiden mutation particularly increases risk of thrombosis to 285:100 000 on the Pill. 3rd generation Pills particularly increase resistance to the body's natural anticoagulant activated protein C (APC), so increasing thrombosis. Those starting the Pill should be counselled that the Pill does increase the risk of thrombosis, but is still a rare event.

The Pill and travel: If immobile for >5h, the *BNF* recommends mid journey exercises ± support stockings.²

1 2000 *Drug Ther Bul* 38 2 *BNF* 2002 i 7.3.1

Depot and implant hormonal contraception

Depot progesterone ('the injection') is simple, safe and very effective but has suffered from adverse publicity. Two preparations are available: *medroxy-progesterone acetate* (Depo-Provera®) 150mg given deep IM 12-weekly; start during the first 5 days of a cycle (or at 5 days post-partum if bottle feeding; 6 weeks if breast feeding) or *norethisterone enanthate* (Noristerat®) 200mg into gluteus maximus 8-weekly (licensed for short-term use only, but can be given immediately postpartum when Depo-Provera® use can cause very heavy bleeding). Exclude pregnancy biochemically and use condoms for 14 days after late injections (eg if she turns up one week late). They inhibit ovulation and thicken cervical mucus. *CI*: Pregnancy; abnormal undiagnosed vaginal bleeding; acute liver disease; severe cardiac disease.

Advantages of progesterone injections:

- No oestrogen content
- Fewer ovarian cysts
- Reduces PMS (p 16)
- No compliance problems
- Reduced endometriosis
- Secret
- Good when GI disease
- Abolishes menorrhagia eventually
- Protects against ectopics
- Reduces PID (p 50)
- Useful when breast feeding
- No thrombosis risk
- 5-fold protective effect against endometrial carcinoma.

Problems: There may be irregular vaginal bleeding. This usually settles with more prolonged use and amenorrhoea often then supervenes, so encourage perseverance. With Depo-Provera® 33% of women have amenorrhoea after 6-months use; 50% after 12 months, and 60% after 18 months (figures are 14%, 27%, and 33% respectively for Noristerat®). If very heavy bleeding occurs, exclude pregnancy; give injection early (but >4 weeks from previous dose) and give oestrogen if not contraindicated (eg Premarin® 1.25mg/24h po for 21 days or a combined oral contraceptive Pill). Fears of osteoporosis in amenorrhoeic women with low oestrogen levels appear unfounded, but use in those <21 years old may prevent optimal bone mineralization.¹ Other problems include weight gain (up to 2kg in 70% of women) and acne.

Special uses: Depot injections may be particularly useful:

- To cover major surgery. If given when stopping combined Pill 4 weeks before surgery it gives contraceptive cover for the next 8 or 12 weeks.
- Sickle-cell disease (reduces incidence of sickle-cell crises).
- Epileptics if on liver enzyme inducers p 63 TB give Depo-Provera® injection every 10 weeks.
- After vasectomy while awaiting partner's sperm-free ejaculates.
- Where bowel disease may affect oral absorption.

2002 Medline 11945109

There may be some delay in return of ovulation on stopping injections (median delay 10 months) but long-term fertility is not reduced.

Implants Progesterone implants give up to 3 years' contraception with one implantation. Implanon® is a flexible rod containing etonogestrel 68mg which is implanted subdermally into the lower surface of the upper arm. It inhibits ovulation and thickens mucus. Contraceptive effect stops when the implant is removed. <23%² of users become amenorrhoeic after 12 months use. Infrequent bleeding occurs in 50% in the first 3 months use; 30% at six months. Prolonged bleeding affects up to 33% in first 3 months; frequent bleeding affects <10%.² Effective contraception may not occur in overweight women in the 3rd year, so consider earlier changing of implant in these women. Liver enzyme inducers (p 63) reduce efficacy, so use other, eg barrier contraception if they are started.

Sterilization

Sterilization is popular, perhaps because it is reliable, without one having to think about it. ~100,000 women and ~90,000 men are sterilized annually in the UK. ~25% of women rely on sterilization for contraception.

Ideally see both partners and consider the following:

- **Alternative methods.** Do they know about the cap, the PoP, depot progesterone injections, coils, and implants?
- **Consent.** Is it the wish of both partners? Legally only the consent of the partner to be sterilized is required but the agreement of both is desirable.
- **Who should be sterilized?** Does she fear loss of her femininity? Does he see it as being neutered? If the woman is requesting sterilization, is she really wanting a hysterectomy or would benefit from one?
- **Irreversibility.** Reversal is only 50% successful in either sex and tubal surgery increases the risk of subsequent ectopics, so the couple should see sterilization as an irreversible step. The sterilizations most regretted are those carried out on the young and at times of stress, or immediately after pregnancy (termination or delivery). Are their children healthy?
- **If she wishes to be sterilized at Caesarean section,** explain that it will only be done if the baby is normal and healthy.
- **Failure rates**—10-year follow-up of 10,863 USA women gave pregnancy rates of 18.5/1000 procedures,¹ no better than new IUCDs (Copper T 380s®, Mirena®, GyneFIX®) and 10-fold worse than vasectomy. ►Remember: IUCDs are reversible, conveniently mirroring many marriages: but sterilization is for ever. Warn that if pregnancy occurs it is likely to be ectopic (32.9%).
- **Side-effects.** A woman who has been on the Pill for many years may find her periods unacceptably heavy after sterilization.

►Record in the notes: *Knows it's irreversible; lifetime failure rate discussed, eg 1:2000 for vasectomy, and 1:200 for Filshie clip ♀ sterilization: see above.*

Female sterilization The more the tubes are damaged, the lower the failure rate and the more difficult reversal becomes. In the UK, most sterilizations are carried out laparoscopically with general anaesthesia. The commonest UK method used is Filshie clips. The tubes may be clipped, ringed, or diathermied. Alternatively the tubes may be divided and ligated through a mini-laparotomy incision, or clipped via the posterior fornix ('colpotomy'). Woman should continue using effective contraception until the operation. An IUCD should be left in place until the next period in case an already fertilized ovum is present. NB: early reports of post-sterilization excessive menstrual blood loss have not been confirmed.¹

Vasectomy This is a simpler procedure than female sterilization and can be performed on an outpatient basis. The vas deferens is identified at the top of the scrotum, ideally 1.5cm is removed, and the cut ends doubled back before being ligated. Bruising and haematoma are complications. Late pain affects 3% from sperm granulomata, which are less common if thermal cautery (rather than electrical cautery) is used.

The major disadvantage of vasectomy is that it takes up to 3 months before sperm stores are used up. Obtain 2 ejaculates 'negative' for sperm (the first 8 weeks post-op; 2nd 2–4 weeks later) before stopping other methods of contraception. Reversal is most successful if within 10 years of initial operation. SE: Evidence of ↑risk of prostate cancer post-vasectomy is equivocal.² None of the positive trials was on European men. The best trials looking at an association between testicular neoplasia and vasectomy have all proved negative.²

Urinary malfunction

Control of bladder function Continence in women is maintained in the urethra by the external sphincter and pelvic floor muscles maintaining urethral pressure higher than bladder pressure. Micturition occurs when these muscles relax and the bladder detrusor muscle contracts.

Urge incontinence 'If I've got to go, I've got to go **now**. The bladder is 'unstable' with high detrusor muscle activity. It occurs in nulliparous and parous women. Usually no organic cause is found, (rarely, neurogenic).

Stress incontinence Small quantities of urine escape as intra-abdominal pressure rises, eg sneezing. Commoner in parous women; pregnancy, not mode of delivery is the risk factor.¹ Examination may reveal prolapse (p 54) or incontinence (ask her to cough).

Detrusor instability usually presents with *urge* incontinence, but in 25% presents as *stress* symptoms. It is a common cause of incontinence (33% of women; 50% of men, of the uk 3.5 million incontinent population.)²

Management

- Exclude a UTI and glycosuria.
- History and examination will not tell you whether the problem is stress or urge or both: arrange urodynamic studies. See p 71 opposite.
- Urge improves with 'bladder training' (gradually increasing the time interval between voiding). For drug treatments see p 71 opposite.
- Mild stress incontinence responds well to pelvic floor exercises or physiotherapy, eg with weighted vaginal cones—insert the heaviest that can be retained (base up) for 15mins twice daily, graduating to heavier cones (max 100g) as tone improves, to improve muscle tone. A vaginal tampon supports the bladder neck, stopping leaks while playing sport.
- Surgery for severe stress symptoms increases intraurethral pressure (by bringing the upper urethra into the zone of abdominal pressure when previously it was below the pelvic floor) and to reduce prolapse. Operations include: urethroplasty, transabdominal colposuspension. For some, tension free tape inserted around the urethra under local anaesthetic, will cure. Others may be helped by periurethral bulking.³
- In some postmenopausal women urgency, increased frequency, and nocturia may be helped by topical estriol 0.1% cream *pv* used long term.
- In those mixed stress/urge incontinence treat the detrusor instability first as this can be made worse by operations for stress incontinence.

True incontinence Continuous leakage of urine may be due to congenital anatomical abnormalities, eg ectopic ureters, or to acquired problems, eg vesicovaginal fistula due to trauma (usually after abdominal hysterectomy in developed countries, after prolonged labour with vertex presentation in developing countries), malignancy, or radiotherapy. If surgery is not possible, seek the help of the continence adviser.

Interstitial cystitis⁴ Characterized by frequency, urgency, bladder pain, and dyspareunia for >6 months, this condition may be helped by dietary modification, bladder training, and biofeedback techniques, drugs (eg antidepressants), and bladder hydrodistension or instillations. www.ihelp.com

Voiding difficulties

Symptoms

- Poor flow
- Straining to void
- Chronic retention
- Stress incontinence
- Intermittent stream
- Hesitancy
- Overflow incontinence
- Urge incontinence
- Incomplete emptying
- Acute retention
- UTI from residual urine

► Remember faecal impaction as a cause of retention with overflow. Where detrusor weakness is the cause there is incomplete bladder emptying with dribbling overflow incontinence. Voiding difficulties are uncommon in women.

Causes

CNS: These may be suprapontine (eg stroke); due to cord lesions (cord injury, multiple sclerosis); peripheral nerve (prolapsed disc, diabetic or other neuropathy); or reflex, due to pain (eg with herpes infections).

Drugs: Especially epidural anaesthesia; also tricyclics, anticholinergics.

Obstructive: Early oedema after bladder neck repair is a common cause. Others: Uterine prolapse, retroverted gravid uterus, fibroids, ovarian cysts, urethral foreign body, ectopic ureterocele, bladder polyp, or cancer.

Bladder overdistension—eg after epidural for childbirth.

Tests

- MSU to exclude infection.
- Ultrasound for residual urine and bladder wall thickness (>6mm on transvaginal scan associated with detrusor instability).¹
- Cystourethroscopy.
- Uroflowmetry (a rate of <15mL/sec for a volume of >150mL is abnormal). Do this test before any surgery is contemplated.
- Urodynamic studies; subtraction cystometry is a subtraction of intra-abdominal pressure from measured intravesical pressure to give detrusor pressure (intravesical measure is a mix of bladder pressure and intra-abdominal pressure).

Treatment This depends upon the cause. For detrusor instability avoid caffeine (mild diuretic, detrusor stimulant); bladder training to increase interval between voiding. Pelvic floor muscle physiotherapy may help those with stress incontinence symptoms. Antimuscarinic drugs may be effective,² eg oxybutynin, starting with 2.5mg/12h, increasing slowly up to 5mg/6h (per 12h if elderly; SE: dry mouth, blurred vision, nausea, headache, constipation, diarrhoea, abdo pain—SEs are less if modified-release once daily tablets are used—30mg/day of Ditropan XL® may be tolerated; approach this by weekly 5mg jumps). Tolterodine eg 2mg/12h is also effective; with lower side-effect profile. For those with nocturia desmopressin (unlicensed use) may reduce frequency in those resistant to other treatment.²

Acute retention may require catheterization (use a suprapubic catheter if needed for several days). For persistent conditions (eg neurological conditions) self-catheterization techniques may be learned, eg with a Lofric® gel coated catheter.

With detrusor weakness drugs may relax the urethral sphincter or stimulate the detrusor muscle. Alpha blockers eg tamsulosin 400µg/24h relax the bladder neck; diazepam relaxes the sphincter. Operative measures may overcome some of the obstructive causes, eg urethrotomy for distal urethral stenosis (uncommon).

Gynaecological surgery

Hysterectomy *Abdominal hysterectomy* is usually total (uterine body + cervix removed), but may be Wertheim's (extended to include local lymph nodes and a cuff of vagina)—used for malignancy. If subtotal, the cervix is left behind—and may become malignant. At *vaginal hysterectomy* the uterus is brought down through the vagina. Hospital stay is shorter but difficulties may arise at operation if the uterus is very bulky. Lower segment Caesarean section and nulliparity may also impede operation.¹ Healthy ovaries are usually conserved, especially in young women, whichever route the uterus is removed by, unless the hysterectomy is for an oestrogen-dependent tumour. Complication: residual ovary syndrome (pain, deep dyspareunia, premature ovarian failure). *Laparoscopic hysterectomy* was developed for patients in whom problems with open surgery are anticipated (eg extensive adhesions with endometriosis), but most of these patients can have a vaginal hysterectomy—which is much quicker (77min vs 131min), with a similar complication rate. Hospital stays are shorter, but total recovery not quicker.²

Most hysterectomies are performed for menorrhagia or fibroids, and rates vary widely. Femininity and sexuality are bound up with the uterus. *Subtotal hysterectomy* leaves the cervix behind so remember to go on smearing these cervixes. The cervix is left in the belief that orgasm is less disrupted (controversial) but late cervical problems (including bleeding) are relatively common (11%).³ ► Women who are counselled, and make their own decision about surgery, are less likely to have regrets. Operative mortality: ~0.06%. Hysterectomy does not cause ↑ psychological/sexual morbidity compared with ablation (below) at 1yr post-op.

Manchester repair (Fothergill's operation) Pregnancy is still possible after this operation for uterine prolapse. The cervix is amputated and the uterus is supported by shortening the ligaments.

Dilatation and curettage (D&C) The cervix is dilated sufficiently to admit a curette to scrape out a sample of endometrium for histology. D&C is a procedure for diagnosing abnormal bleeding (but outpatient endometrial sampling (p 41) may make D&C unnecessary). Adequate sampling occurs in 75% but in 10% abnormal pathology may be missed.⁴ Evacuation of retained products of conception from the uterus after miscarriage (ERPC), or termination of early pregnancy, are carried out by **dilatation and suction**.

Hysteroscopy As an outpatient alternative to D&C, a hysteroscope can be inserted through the cervix into the uterus to visualize the endometrium. 'Blind' samples may then be taken using a sampler.

Hysteroscopic endometrial ablation by laser, diathermy, or other ablative method⁵ (under GA or spinal ± paracervical block) reduces bleeding by achieving a deliberate Asherman's syndrome (p 12); as an alternative to hysterectomy, it has fewer complications. Endometrium may be thinned pre-op by leuprolerin or danazol. By 4 months 10% have menorrhagia again. Complications: (major in 4% requiring further operation in 1% haemorrhage; infection (eg late necrotizing granulomatous endometritis); uterine perforation; haematometra, vesicovaginal fistula, fluid overload from irrigation fluid can cause ↑BP, ↓Na⁺, pulmonary oedema, CNS symptoms, and haemolysis. Some endometrium remains in the majority (so give progesterone containing HRT later, if needed). Pregnancy and endometrial cancer can still occur after ablation. See p 14.

Laparoscopy The laparoscope is inserted sub-umbilically. Instrument

are inserted through ports in the *iliac fossae*. Occasionally a Verres needle is placed suprapubically to insufflate and aid manipulation of organs. This procedure allows visualization of the pelvic organs and is used for diagnosis of pelvic pain and ectopic pregnancy. The patient is spared a full laparotomy unless needed for treatment. A 'lap and dye' demonstrates tubal patency. Sterilization and hysterectomy may be laparoscopically carried out, and ectopic pregnancies treated.

Colporrhaphy or '**repair**' The lack of support from the vaginal wall in cases of prolapse is rectified by excising a piece of the redundant mucosa and plicating levator ani. It is *not* an operation to correct urinary incontinence. The operation may be combined with Manchester repair or vaginal hysterectomy. The more mucosa is removed, the tighter the vagina. Enquire before surgery if she is sexually active. Catheterization circumvents post-operative retention of urine.

Cone biopsy A cone of tissue (point inwards) is cut out around the external cervical os, using knife or laser. This removes neoplastic tissue for histology, and may be curative. Complications: (immediate) bleeding, (long-term) cervical stenosis or incompetence.

Pelvic exenteration Consider this option when initial surgery fails to control neoplasia of the cervix, vulva, or vagina. It involves removal of the pelvic organs—ie ultra-radical surgery, which should only be contemplated if there is a chance of cure. Do your best to establish whether disease has spread to the pelvic sidewall or nodes, eg with MRI or CT scans and intra-operative biopsy with frozen section: if so, exenteration is probably not worthwhile. Only ~20% of possible candidates for surgery meet this criterion: in addition the patient should be quite fit, and ideally have a supportive partner. We know that palliative exenteration in those with unresectable disease is not worthwhile. 5-yr survival: ~50%. Operative mortality: ~5%. Complications: GI obstruction/fistulae; urinary fistulae. Remember to give full pre-operative counselling about colostomies, and sexual function (refashioning of the vagina may be possible).

Chronic pelvic pain

This can be a cause of much misery to women of reproductive age. The history is usually of longstanding pelvic pain with secondary dysmenorrhoea and deep dyspareunia. The pain may cause, or be exacerbated by, emotional problems. She may be depressed.

Laparoscopy may reveal a likely cause: chronic pelvic infection, endometriosis, adenomyosis, adhesions, or congested pelvic veins. If it does not (or if all gynaecological causes have been surgically removed) the cause may be gastrointestinal: consider irritable bowel syndrome (*OHCM* p 236).

Pelvic congestion Lax pelvic veins become painfully congested with blood. The pain is worse when she is standing, walking (gravity fills the veins), and premenstrually. It is typically variable in site and intensity and there is unpleasant postcoital ache. She is maximally tender to deep palpation over the ovaries. The vagina and cervix may appear blue due to congestion and there may be associated posterior leg varicosities. The dilated veins may be demonstrated by venography or laparoscopically.

Relief may be difficult, though explanation helps ('pelvic migraine'). Medroxyprogesterone 50mg/24h po for 3 months reduces pain (see amenorrhoea, weight gain, bloating); migraine remedies (*OHCM* p 332) and relaxation may be tried. When symptoms are very severe bilateral ovarian vein ligation, or embolization radiologically, may cure, as may hysterectomy with bilateral salpingo-oophorectomy with hormone replacement therapy post-operatively.

Mittelschmerz This is mid-cycle menstrual pain which may occur in teenagers and older women around the time of ovulation—from the German 'mittel' (=middle) and 'Schmerz' (=pain).

Dyspareunia

This means pain during intercourse. There may be a vicious circle in which anticipation of pain leads to tense muscles and lack of lubrication, and so to further pain. ► *The patient may not volunteer the problem so ask about intercourse.* Her attitude to pelvic examination may tell you as much as the examination itself. Ask her to show you where the problem is. If the problem is actually vaginismus do not insist on examination and consider counselling and sex therapy (p 380). Was there 'female circumcision' (p 6)?

Dyspareunia may be superficial (introital)—eg from infection so look for ulceration and discharge. Is she dry? If so is the problem oestrogen deficiency (p 18) or lack of sexual stimulation? Has she had a recent post-partum perineal repair? A suture or scar can cause well-localized pain which is cured by removing the suture and injection of local anaesthetic. If the introitus has been rendered too narrow, she may need surgery.

Deep dyspareunia is felt internally (deep inside). It is associated with endometriosis and pelvic sepsis; treat the cause if possible. Ovaries lying in the rectovaginal pouch (or after hysterectomy), may be subject to coital thrusts; try other positions or ventrosuspension if a 'cure' can be obtained with trial use of a Hodge pessary.

Dermatographism is a rare cause of dyspareunia: look for itchy vulva wheals some minutes after calibrated dermatographometer application. It can occur on any surface. It is the commonest physical cause of urticaria and the clue to its presence is linear wheals with a surrounding bright red flare (but no angio-oedema) elsewhere on the skin, in response to a firm stroke. Cause is unknown. Relief of dyspareunia in these cases has been achieved by 2% epinephrine (adrenaline) cream, and cetirizine 10mg/24h po.

Ovarian hyperstimulation¹

Sudden multiplication of follicles in response to drugs, especially when gonadotrophins are used in assisted reproduction, gives diffuse lower abdominal pain and cystic ovarian enlargement on palpation. The incidence for severe cases is 0.5–2% in women having ovarian stimulation (mild cases occur in 22–33% of treatment cycles)². It is commoner in conception cycles. Women with polycystic ovarian syndrome are at particular risk.² Associated complications are ascites, reduced intravascular volume, pulmonary effusions and (rare) pericardial effusions. Thromboembolic complications may ensue (especially in upper limb and cerebral vessels).

Presentation Abdominal discomfort, nausea, vomiting, and abdominal distension \pm dyspnoea. Presentation is usually 3–7 days after hCG administration, or 12–17 days, if pregnancy has ensued.

Prevention³ In those in whom hyperstimulation is suspected (peak serum oestradiol $>6000\text{pg/mL}$ and >30 follicles stimulated), the surest way to avoid hyperstimulation is to avoid administration of human chorionic gonadotrophin (hCG) so the stimulated follicles do not ovulate, thereby cancelling the cycle of treatment (or discontinue gonadotrophins and delay hCG injections until the serum oestradiol returns to 'safe' levels.) Some selectively cryopreserve embryos for later transfer.

Risk factors

- Age $<35\text{yrs}$
- Asthenic habitus
- Pregnancy
- Luteal phase hCG stimulation
- Polycystic ovarian syndrome
- Serum oestradiol $>4000\text{pg/mL}$
- Multiple follicles (>35)

Management Admit to hospital. Give adequate analgesia (opiates if necessary). Check FBC, U&E, creatinine, LFTs, and coagulation profile. Do CXR if chest pathology suspected; blood gases if tachypnoeic or dyspnoeic. Chart urine output (if oliguria suspected catheterize and measure fluid output hourly). Ovarian size is a guide to severity unless oocytes have been harvested.

Hyperstimulation is severe if haematocrit is $>45\%$, $\text{wcc} >15 \times 10^9/\text{L}$, there is massive ascites, oliguria, mild renal, or liver dysfunction. Put up ivi, and consider cvp monitoring. Fluid replacement may be required, eg 500mL albumin over 2 hours. Paracentesis of ascites can improve renal function. To avoid thromboembolic complications use antiembolic stockings, encourage leg mobility, and consider use of subcutaneous heparin.

The situation is *critical* if the haematocrit is $>55\%$ and $\text{wcc} >25 \times 10^9/\text{L}$, there is tense ascites and renal failure, thromboembolic phenomena, and acute respiratory distress syndrome (OHCM p 176). Admit to ITU. Symptomatic pleural effusions may need drainage. Use antiembolic measures as above. Pay meticulous attention to fluid balance. Aim to maintain intake at 3L/24h using alternating normal and dextrose saline (but beware hyponatraemia).

The severity of effusions determines the time to recovery. In non-conception cycles with little ascites there will be resolution with menstruation. In conception cycles cysts may persist for weeks. Resolution usually occurs, but exceptionally termination of pregnancy may be needed. Deaths have been reported but are extremely rare ($\sim 1:500,000$ stimulated cycles).²

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Sources *Cochrane childbirth & pregnancy database*: Royal College of Obstetricians and Gynaecologists; *BMJ*.

*The term *pregnancy-induced hypertension with proteinuria* is tending to replace the term *pre-eclampsia*. We have not followed this trend as to do so obscures the vital fact about pre-eclampsia: it may lead on to eclampsia. We favour *pre-eclampsia* because it is short and sends the shadow of a shiver down our spines, being a reminder of how dangerous it can be.

Relevant pages in other chapters: Neonatology p 230-46; breast feeding p 176-8; rhesus disease p 242; ectopic pregnancy p 24; abortion and termination p 20-2; trophoblastic disease p 26; fibroids in pregnancy p 39; neonatal examination p 231; preterm and light-for-dates babies p 180; Cystic fibrosis p 193; varicella (chickenpox) in pregnancy p 216; parvovirus B19 p 214; postnatal depression p 408.

The essence of reproductive health

Pregnancy is a risky affair, not only for babies, but also for mothers. The textbook causes of maternal mortality in the UK are pulmonary embolism, eclampsia, haemorrhage, and infection—with all the other causes being rare. But if an obstetrician could be granted one wish, it would not be to abolish these; rather it would be to make every pregnancy *planned and desired by the mother*. Worldwide, a woman dies every minute from the effects of pregnancy, and most of these women never wanted to be pregnant in the first place—but either had no means of contraception, or were without the skills, authority, and self-confidence to negotiate with their partners.¹ So the real killers are poverty, ignorance, and the unwieldy desires of men, and the real solutions entail literacy, economic growth, and an equality of dialogue between the sexes. Any obstetric or governmental initiatives in reproductive health which do not recognize these facts are doomed.²

School-based sex education This can be effective, if linked to easy access to contraceptive services. This is the conclusion of a meta-analysis, taking into account cohort studies (if meta-analyses confine themselves to the 15-or so randomized studies, no benefit could be shown).³ It may be necessary to foster a knowledge-sharing, skill-promoting environment that is part of a continuing process, and not a 'one-off' affair—for educational programmes to work.

Adolescent pregnancy rates: USA: 116/1000; UK: 57/1000; Canada: 50/1000. In the UK, 2,123,614 babies were born in 1997–9 (live & stillbirths), and 564,100 pregnancies were legally terminated.

Definitions *Gravidity* refers to the number of pregnancies that a woman has had (to any stage).

Parity refers to pregnancies that resulted in delivery beyond 28 weeks' gestation. An example of the shorthand way of expressing pregnancies before and after 28 weeks is: para 2+1. This means that she has had 2 pregnancies beyond 28 completed weeks' gestation, and 1 which terminated prior to 28 weeks. If she is not pregnant at the time of describing her she is gravida 3, but if she is pregnant now she is gravida 4. Twins present a problem as there is controversy as to whether they count as 1 for both parity and gravidity or should count as 2 for parity.

It is unclear whether the cut-off point in these definitions should now be 24 weeks, to harmonize with the new definition of stillbirth (p 136). In general, aim to use proper English rather than the shorthand described above, which is open to ambiguity. For example, when presenting a patient try something like: 'Mrs Cottard is a 32-year-old lady who is 15 weeks into her 4th pregnancy; the 3rd ended in a miscarriage at 17 weeks, and the others came to term with normal deliveries of children who are now 2 & 8.' The bald statement 'Para 2+1' is ambiguous, incomprehensible to the patient, and misses the point that the patient is now approaching the time when she lost her last baby.

The placenta

The placenta is the organ of respiration, nutrition, and excretion for the fetus. It produces hormones for maternal wellbeing and immunologically protects the fetus by preventing rejection and allowing the passage of IgG antibodies from the mother.

Development At term the placenta weighs $\frac{1}{7}$ th the weight of the baby. It has a blood flow of 600mL/min. The placenta changes throughout pregnancy as calcium is deposited in the villi and fibrin on them. Excess fibrin may be deposited in diabetes and rhesus disease, so ↓ fetal nutrients.

Placental types *Battledore* insertion is where the umbilical cord inserts into the side of the placenta. *Velamentous* insertion (1%) is where the umbilical vessels pass within the membranes before insertion. If these vessels break (as in vasa praevia) it is fetal blood that is lost. *Placenta succenturia*: (5%) There is a separate (succenturiate) lobe away from the main placenta which may fail to separate normally and cause a PPH or puerperal sepsis. *Placenta membranacea* (1/3000) is a thin placenta all around the baby. As some is in the lower segment it predisposes to APH. It may fail to separate in the third stage of labour. *Placenta accreta*: There is abnormal adherence of all or part of the placenta to the uterus, termed *placenta increta* where there is placental infiltration of the myometrium or *placenta percreta* if penetration reaches the serosa. These latter 3 types predispose to PPH and may necessitate hysterectomy.

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Placenta praevia The placenta lies in the lower uterine segment. It is found in ~0.5% of pregnancies.¹ Risks are of significant haemorrhage by mother and fetus. **Associations:** Large placenta (eg twins); uterine abnormalities and fibroids; uterine damage, eg multiparity; former surgery (caesarean section, myomectomy); past infection. Ultrasound at <24 weeks gestation shows a low-lying placenta in 28% but lower segment development later in pregnancy results in only 3% being low-lying at term. Transvaginal ultrasound is superior to transabdominal for localizing placenta accurately, and, if combined with Doppler, diagnose vasa praevia and placenta accreta. It has not been shown to increase bleeding.²

Major (old III and IV degrees) with placenta covering the internal os requires caesarean section for delivery. Minor (old I and II) where the placenta is in the lower segment but not across the internal os: aim for normal delivery. Presentation may be as APH (separation of the placenta as the lower segment stretches causes bleeding) or as failure for the head to engage with high presenting part. Problems are with bleeding and with mode of delivery as the placenta obstructs the os and may shear off during labour, or may be accreta (5%), especially after previous caesarean section (>24%). Caesarean section should be consultant-performed or supervised. The rule of admitting those with major placenta praevia at ≤35 weeks' gestation so that immediate help is available, is controversial, and not practiced by many UK units.³

After delivery Examine placenta for abnormalities (clots, infarcts, amnion nodosum—p 120, vasa praevia, single umbilical artery). Weigh the placenta (weight >25% of the baby suggests congenital nephrotic syndrome). Blood may be taken from the cord for Hb, Coombs test, LFTs, and blood group (eg for rhesus disease), or for infection screens, if needed.

1 Variable, eg 0.38% in singletons, see M Katz 2001 *J Mat Fet Med* 10 414 [11798453]

2 K Oyelese 1999 *Obs Gy Sur* 54 138 [9950006] 3 Z Penn 2001 *BMJ* ii 165 [11484722]

Plasma biochemistry in pregnancy^{1,2,3}

Centile	Non-pregnant		Trimester 1		Trimester 2		Trimester 3	
	2.5	97.5	2.5	97.5	2.5	97.5	2.5	97.5
Na ⁺ mmol/L	138	146	135	141	132	140	133	141
Ca ²⁺ mmol/L	2	2.6	2.3	2.5	2.2	2.2	2.2	2.5
*corrected	2.3	2.6	2.25	2.57	2.3	2.5	2.3	2.59
Albumin g/L	44	50	39	49	36	44	33	41
FreeT ₄ pmol/L	9	23	10	24	9	19	7	17

Other plasma reference intervals (not analysed by trimester)

	Non-pregnant	Pregnant
Alkaline phosphatase	3–300iu/L	up to 450iu/L **
Bicarbonate mmol/L	24–30	20–25
Creatinine µmol/L	70–150	24–68
Urea mmol/L	2.5–6.7	2–4.2
Urate µmol/L	150–390	100–270

*Calcium corrected for plasma albumin (see *OHCM* p 696).

**Occasionally very much higher in apparently normal pregnancies.⁴

C-reactive protein does not change much in pregnancy.

TSH may be low in the first half of a normal pregnancy (suppressed by hCG); for other thyroid changes see above and p 157.

Protein S falls in pregnancy, so protein S deficiency is difficult to diagnose. Activated protein C (aPC) resistance is found in 40% of pregnancies so special tests are needed when looking for this. Genotyping for Factor V Leiden and prothrombin G20210A are unaffected by pregnancy.

Physiological changes in pregnancy

Hormonal changes *Progesterone*, synthesized by the corpus luteum until 35 post-conception days and by the placenta mainly thereafter, decreases smooth muscle excitability (uterus, gut, ureters) and raises body temperature. *Oestrogens* (90% oestriol) increase breast and nipple growth, water retention and protein synthesis. The maternal thyroid often enlarges due to increased colloid production. Thyroxine levels, see p 157. Pituitary secretion of *prolactin* rises throughout pregnancy. Maternal *cortisol* output is increased but unbound levels remain constant.

Genital changes The 100g non-pregnant uterus weighs 1100g by term. Muscle hyperplasia occurs up to 20 weeks, with stretching after that. The cervix may develop ectropion ('erosions'). Late in pregnancy cervical collagen reduces. Vaginal discharge increases due to cervical ectopy, cell desquamation, and ↑ mucus production from a vasocongested vagina.

Haemodynamic changes *Blood*: From 10 weeks the plasma volume rises until 32 weeks when it is 3.8 litres (50% >non-pregnant). Red cell volume rises from 1.4 litres when non-pregnant to 1.64 litres at term if iron supplements not taken (↑18%), or 1.8 litres at term (↑30%) if supplements are taken—hence Hb falls due to dilution (physiological 'anaemia'). wcc (mean $10.5 \times 10^9/L$), platelets, esr (up 4-fold), cholesterol, β -globulin, and fibrinogen are raised. Albumin and gamma-globulin fall.

Cardiovascular: Cardiac output rises from 5 litres/min to 6.5–7 litres/min in the first 10 weeks by increasing stroke volume (10%) and pulse rate (by ~15 beats/min). Peripheral resistance falls (due to hormonal changes). BP, particularly diastolic, falls during the first and second trimesters by 10–20mmHg, then rises to non-pregnant levels by term. With increased venous distensibility, and raised venous pressure (as occurs with any pelvic mass), varicose veins may form. Vasodilatation and hypotension stimulates renin and angiotensin release—an important feature of BP regulation in pregnancy.

Other changes Ventilation increases 40% (tidal volume rises from 500 to 700mL), the increased depth of breath being a progesterone effect. O_2 consumption increases only 20%. Breathlessness is common as maternal P_aCO_2 is set lower to allow the fetus to offload CO_2 . Gut motility is reduced, resulting in constipation, delayed gastric emptying, and with a lax cardiac sphincter, heartburn. Renal size increases by ~1cm in length during pregnancy.

Frequency of micturition emerges early (glomerular filtration rate ↑ by 60%), later from bladder pressure by the fetal head. The bladder muscle is lax but residual urine after micturition is not normally present. Skin pigmentation (eg in linea nigra, nipples, or as chloasma—brown patches of pigmentation seen especially on the face), palmar erythema, spider naevi, and striae are common. Hair shedding from the head is reduced in pregnancy but the extra hairs are shed in the puerperium.

Pregnancy tests Positive eg from two weeks post-conception (or from the first day of the first missed period), until ~20 weeks of pregnancy, they remain positive for ~5 days after abortion or fetal death. Otherwise, the false +ve rate is low. They detect the β -subunit of human chorionic gonadotrophin in early morning urine, so are positive in trophoblastic disease (p 26).

Thrombotic sequelae of physiological changes in pregnancy

Routine antenatal and postnatal care needs to take into account the fact that pregnancy is a hypercoagulable state. In order to minimize risk of thromboembolism, thromboprophylaxis is now routinely recommended in certain circumstances. For the context of caesarean section, see p 133.

Thromboprophylaxis after vaginal delivery:¹

Low risk: Those with uncomplicated pregnancy. To minimize risks, early mobilization and avoiding dehydration is usually sufficient.

Moderate risk: Women with any **two** of characteristics below:

- Age >35y
- Parity ≥4
- Labour >12h
- Pre-eclampsia
- Current infection
- Obesity (booking weight >80kg)
- Gross varicose veins
- Immobility for ≥4 days prior to labour
- Mid cavity or rotational forceps
- Major current medical illness

Treat with enoxaparin 40mg/24h sc starting within 6h of delivery (but ≥4h after removal of an epidural catheter). Continue until hospital discharge—review at 5 days if still an inpatient. If heparin is contraindicated use TED elastic compression stockings (TED=transverse elastic graduated).

High risk: Women with **one** of:

- 4 or more risk factors listed above
- Extended pelvic or abdominal surgery
- Family history of deep vein thrombosis, pulmonary embolus, thrombophilia, or personal history of past thromboembolism
- Paralysis of lower limbs
- Antiphospholipid antibodies or lupus anticoagulant

Treat with enoxaparin as above and to wear elastic stockings. If known thrombophilia or past history of thromboembolism treat for 6 weeks. Seek specialist advice for those with a family history of thrombophilia.

Thromboprophylaxis for women flying in pregnancy²

This is for any gestation and up to 6 weeks postpartum.

- All women should do isometric calf exercises in flight, walk around the cabin if possible, drink plenty of soft drinks/water, avoiding alcohol & coffee.
- All on long haul (≥4 hour) flights should wear knee length graduated compression stockings.
- Women with additional risk factors eg weight >100kg, body mass index at booking >30; multiple pregnancy; thrombophilia; those with a past or family history of thromboembolism should wear stockings even on short haul flights; on long haul flights they should consider low dose heparin, eg 40mg enoxaparin or 5000u dalteparin sc pre-flight and on the day after travel; or if heparin is unsuitable, use aspirin 75mg/24h po for 3 days pre-travel and on the day of travel.

Normal labour

From the 1st trimester, the uterus has Braxton-Hicks contractions¹ (ie non-painful 'practice' contractions, eg to ≤ 15 mmHg pressure; in labour pressure is ~ 60 mmHg). They are commonest after 36 weeks.

Normal labour is that occurring after 37 weeks' gestation. It should result in the spontaneous vaginal delivery of the baby within 24h of the onset of regular spontaneous contractions. It is often heralded by a 'show', ie a plug of cervical mucus and a little blood as the membranes strip from the os. The membranes may then rupture.

The first stage of labour is the time from the onset of regular contractions until the cervix is fully dilated (no cervix felt around the head). The cervix initially effaces (becomes shorter and softer) before it dilates. A satisfactory rate of dilatation from 3cm dilated is 1cm/h. The first stage generally takes up to 12h in a primip, and 7.5h in a multip. During the first stage check maternal pulse, BP, and T^o half-hourly; assess the contractions every 15min, their strength (you should not be able to indent the uterus with the fingers during a contraction) and their frequency (ideally 3–4 per 10min, lasting up to 1min). Carry out vaginal examination eg every 4h to assess the degree of cervical dilatation, the position and the station of the head (measured in cm above the ischial spines) and note the degree of moulding (p 86). Note the state of the liquor (see p 128). Test maternal urine 4-hourly for ketones and protein. If the mother becomes ketotic set up an ivi and give her 10% dextrose.* Measure the fetal heart rate (if not being continuously monitored) every 15min. Note the rate before, during, and immediately after a contraction.

The second stage is the time from complete cervical dilatation until the baby is born (see *Movement of head in labour*, p 86).

The mother has an urge to push and uses abdominal muscles and the Valsalva manoeuvre to help move the baby. As the head descends, the perineum stretches and the anus gapes. Normal time for second stage is 45–120min in a primip, and 15–45min in a multip. Prevent a precipitate delivery (and so intracranial bleeding) by pressure over the perineum.

Delay in clamping the cord for 30sec and holding the baby 20cm below the introitus results in higher haematocrit levels, so reducing transfusion and oxygen supplement requirements in premature babies.

The third stage is delivery of the placenta. As the uterus contracts to a <24-week size after the baby is born, the placenta separates from the uterus through the spongy layer of the decidua basalis. It then buckles and a small amount of retroplacental haemorrhage aids its removal.

Signs of separation: Cord lengthening → rush of blood (retroplacental haemorrhage) *per vaginam* → uterus rises → uterine contracts in the abdomen (felt with hand as a globular mass).

Routine use of syntometrine (ergometrine maleate 500µg IM + oxytocin 5u IM) as the anterior shoulder is born has decreased third stage time (to ~ 5 min), and decreased the incidence of PPH, but may cause problems for undiagnosed twins. It can precipitate myocardial infarction and is contraindicated in those with ischaemic heart disease and familial hypercholesterolaemia.² Examine the placenta to check it is complete.

*Beware giving large amounts of dextrose in ivis with no sodium—may cause hyponatraemia and other neonatal problems. [@7657008] [@8886680] [@2040241] [@6487563]

1 W Schmidt [@6924911] 2 H Mousa 2000 *BJOG* 107 939 [@10901569]



(1)
1st stage of labour. The cervix dilates. After full dilatation the head flexes further and descends further into the pelvis.



(2)
During the early second stage the head rotates at the level of the ischial spine so the occiput lies in the anterior part of pelvis. In late second stage the head breaches the vulval ring (crowning) and the perineum stretches over the head.



(3)
The head is born. The shoulders still lie transversely in the midpelvis.



(4)
Birth of the anterior shoulder. The shoulders rotate to lie in the anteroposterior diameter of the pelvic outlet. The head rotates externally, 'restitutes', to its direction at onset of labour. Downward and backward traction of the head by the birth attendant aids delivery of the anterior shoulder.



(5)
Birth of the posterior shoulder is aided by lifting the head upwards whilst maintaining traction.

Abdominal palpation

The uterus occupies the pelvis and cannot be felt *per abdomen* until about 12 weeks' gestation. By 16 weeks it lies about half way between the symphysis pubis and the umbilicus. By 20–24 weeks it has reached the umbilicus. In a primigravida, the fundus lies under the ribs by 36 weeks. At term the uterus tends to lie a little lower than at 36 weeks due to the head descending into the pelvis. Some attendants prefer to measure the symphysis fundal height (SFH) in cm from the symphysis pubis (after voiding urine). From 16 weeks the SFH increases ~1cm/week. As a rule of thumb: at 16–26 weeks the SFH (cm) \approx dates (in weeks)

26–36 weeks

the SFH (cm) \pm 2cm \approx dates

36 weeks to term

the SFH (cm) \pm 3cm \approx dates.

SFH is used as a rough guide to find babies small for gestational age (p 104)

Suspect this if the measurement lies >1–2cm outside these ranges given above. NB: more false positives will occur with the simpler rule of weeks of gestation = cm from pubic symphysis to fundus.^{1,2}

Other reasons for discrepancy between fundal height and dates: • Inaccurate menstrual history • Multiple gestation • Fibroids • Polyhydramnios • Adnexal mass • Maternal size • Hydatidiform mole.

On inspecting the abdomen note any scars from previous operations. Caesarean section scars are usually Pfannenstiel ('bikini-line'). Laparoscopy scars are just below and parallel to the umbilicus or in the left upper quadrant (Palmer's point). It is common to see a line of pigmentation, the linea nigra, extending in the midline from pubic hair to umbilicus. This darkens during the first trimester (the first 13 weeks).

Palpating the abdomen Measure the SFH and listen to the fetal heart. After 32 weeks palpate laterally to assess the lie, then bimanually palpate over the lower uterine pole for presentation and degree of engagement. Pawlik's grip (examining the lower pole of the uterus between the thumb and index fingers of the right hand) can also be used for assessing the degree of engagement. Watch the patient's face during palpation and stop if it causes pain. Obesity, polyhydramnios and tense muscles make it difficult to feel the fetus. Midwives are skilled at palpation, so ask them if you need help.

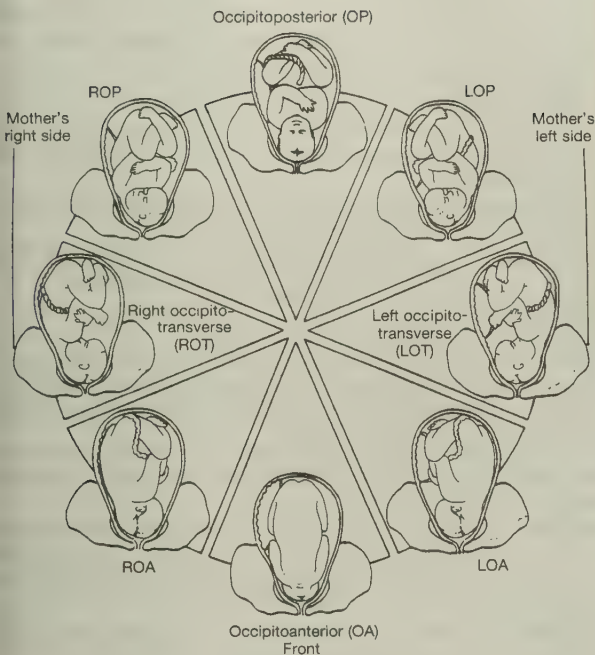
It is important to determine the *number of fetuses* (p 122), the *lie* (longitudinal, oblique, or transverse), the *presentation* (cephalic or breech) and the *engagement*. Ultrasound is useful here. Note the amount of liquor present, the apparent size of the fetus, and any contractions or fetal movements seen or felt.

Auscultation The fetal heart may be heard by Doppler ultrasound from ~12 weeks and with a Pinard stethoscope from ~24 weeks.

Engagement The level of the head is assessed in 2 ways—engagement or fifths palpable abdominally. Engagement entails passage of the maximum diameter of the presenting part through the pelvic inlet. Fifths palpable abdominally states what you can feel, and makes no degree of judgment on degree of engagement of the head. In primigravida the head is usually entering the pelvis by 37 weeks, otherwise causes must be excluded (eg placenta praevia or fetal abnormality). In multips the head may not enter the pelvis until the onset of labour.

1 www.vh.org/Providers/ClinRef/FPHandbook/Chapter14/02-14.html

2 www.arcmesa.com/pdf/emerdeli/emerdeli_chap2.htm



Position

Occipitoanterior

Back easily felt
Limbs not easily felt
Shoulder lies 2cm from
midline on opposite
side from back
Back from midline = 2–3cm 6–8cm

Occipitolateral

Back can be felt
Limbs lateral
Midline shoulder

Occipitoposterior

Back not felt
Limbs anterior
Shoulder 6–8cm
lateral, same
side as back
≥10cm

►The fetal heart is best heard over its back (left scapula).

Pelvis and head

The ideal pelvis This has a rounded brim, a shallow cavity, non-prominent ischial spines, a curved sacrum with large sciatic notches and sacrospinous ligaments >3.5cm long. The angle of the brim is 55° to the horizontal, the AP diameter at least 12cm and transverse diameter at least 13.5cm. The subpubic arch should be rounded and the intertuberosous distance at least 10cm. A *clinically favourable* pelvis is one where the sacral promontory cannot be felt, the ischial spines are not prominent, the subpubic arch and base of supraspinous ligaments both accept 2 fingers and the intertuberosous diameter accepts 4 knuckles when the woman is examined.

The true pelvis Anteriorly there is the symphysis pubis (3.5cm long) and posteriorly the sacrum (12cm long).

Zone of inlet: Boundaries: Anteriorly lies the upper border of the pubis posteriorly the sacral promontory, laterally the ileopectineal line. Transverse diameter 13.5cm; AP diameter 11.5cm.

Zone of cavity: This is the most roomy zone. It is almost round. Transverse diameter 13.5cm; AP diameter 12.5cm.

Zone of mid-pelvis: Boundaries: Anteriorly, the apex of the pubic arch posteriorly the tip of the sacrum, laterally the ischial spines (the desirable distance between the spines is >10.5cm). Ovoid in shape, it is the narrowest part.

Zone of outlet: The pubic arch is the anterior border (desirable angle > 85°). Laterally lie the sacrotuberous ligaments and ischial tuberosities posteriorly the coccyx.

Head terms The *bregma* is the anterior fontanelle. The *brow* lies between the bregma and the root of the nose. The *face* lies below the root of the nose and supraorbital ridges. The *occiput* lies behind the posterior fontanelle. The *vertex* is the area between the fontanelles and the parietal eminences.

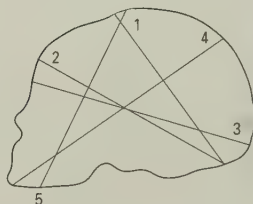
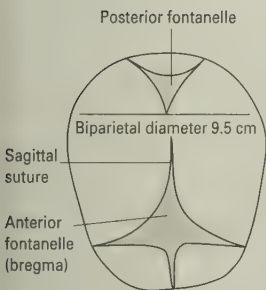
Moulding The frontal bones can slip under the parietal bones which can slip under the occipital bone so reducing biparietal diameter. The degree of overlap may be assessed vaginally.

Presentation:

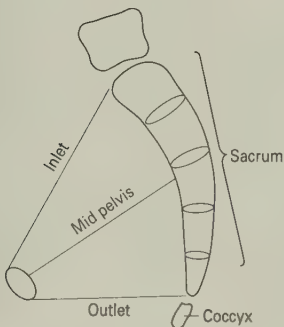
	Relevant diameter presenting:	
Flexed vertex	suboccipitobregmatic	9.5cm
Partially deflexed vertex	suboccipitofrontal	10.5cm
Deflexed vertex	occipitofrontal	11.5cm
Brow	mentovertical	13cm
Face	submentobregmatic	9.5cm

Movement of the head in labour (normal vertex presentation)

- 1 Descent with increased flexion as the head enters the cavity. The sagittal suture lies in the transverse diameter of the brim.
- 2 Internal rotation occurs at the level of the ischial spines due to the grooved gutter of the levator muscles. Head flexion increases. (The head rotates 90° if occipitolateral position, 45° if occipitoanterior, 135° if occipitoposterior.)
- 3 Disengagement by extension as the head comes out of the vulva.
- 4 Restitution: as the shoulders are rotated by the levators until the bisacromial diameter is anteroposterior, the head externally rotates the same amount as before but in opposite direction.
- 5 Delivery of anterior shoulder by lateral flexion of trunk posteriorly.
- 6 Delivery of posterior shoulder by lateral flexion of trunk anteriorly.
- 7 Delivery of buttocks and legs.



- 1 Suboccipitobregmatic 9.5 cm flexed vertex presentation
- 2 Suboccipitofrontal 10.5 cm partially deflexed vertex
- 3 Occipitofrontal 11.5 cm deflexed vertex
- 4 Mentovertical 13 cm brow
- 5 Submentobregmatic 9.5 cm face



Dystocia

Dystocia is difficulty in labour. There may be problems with *the passenger* (large baby, see impacted shoulders=shoulder dystocia p 128, or an abnormal presentation), *the passages* (for ideal pelvis, see p 86—note that cervical dystocia may be a problem after biopsy of the cervix) or of *propulsion* (the uterine powers). Cephalo–pelvic disproportion results if diameters are unfavourable (p 86).

The pelvis The ideal pelvis has a round brim (ie gynaecoid), but 15% of women have a long oval brim (anthropoid). A very flat brim is less favourable (platypoid); occurring in 5% of women over 152cm (5ft), it occurs in 30% of women <152cm. Spinal scoliosis, kyphosis, sacralization of the L5 vertebra, spondylolisthesis and pelvic fractures may all affect pelvic anatomy. Rickets and polio were formerly important causes of pelvic problems. Suspect pelvic contraction if the head is not engaged by 37 weeks in a Caucasian primip (after excluding placenta praevia).

The presentation Cephalic presentations are less favourable the less flexed the head. Transverse lie and brow presentations will always need caesarean section: face and *op* (p 126) presentations may deliver vaginally but are more likely to fail to progress. Breech presentation is particularly unfavourable if the fetus >3.5kg.

The uterine powers Contractions start in the fundus and propagate downwards. The intensity and duration of contractions are greatest at the fundus, but the contraction reaches its peak in all parts of the uterus simultaneously. Normal contractions occur at a rate of 3 per 10min, they should last up to 75sec, the contraction peak usually measures 30–60mmHg, and the resting uterine tone between them should be 10–15mmHg. Uterine muscle has the property of retraction. The shortening of the muscle fibres encourages cervical dilatation.

Uterine dysfunction Contractions may be hypotonic (low resting tone, low contraction peaks) or they may be normotonic but occur too infrequently. These dysfunctions can be corrected by augmentation with oxytocin (p 112). Whenever oxytocin is used discuss with a senior obstetrician. Pain and fear cause release of catecholamines which can inhibit uterine activity. Thus adequate analgesia is needed (p 114) and may speed the progress of labour.

Cervical dystocia Failure of cervical dilatation may be due to previous trauma, repair, cone biopsy, and cauterization. It is difficult to distinguish from failure to dilate due to uterine dysfunction though the latter should respond to oxytocin (note the important difference between primips and multips, p 112). The treatment for cervical dystocia is delivery by caesarean section.

Consequences of prolonged labour Neonatal mortality rises with prolonged labour as does maternal morbidity (especially infection). With modern management of labour, careful monitoring of progress in labour (p 112) takes place to diagnose delay early, and treat it as necessary, to prevent prolonged labour occurring.

►When there is dystocia, ask 'is safe vaginal delivery possible?'

Minor symptoms of pregnancy

► Before prescribing any drug, think—Is it necessary. Is it safe? Consult Data-sheets ± a national teratology information service (0191 232 1525^{uk}).

Symptoms and signs in the first 10 weeks: Early symptoms are amenorrhoea, nausea, vomiting, and bladder irritability. Breasts engorge, nipples enlarge (darken at 12 weeks), Montgomery's tubercles (sebaceous glands on nipples) become prominent. Vulval vascularity increases and the cervix softens and looks bluish (4 weeks). At 6–10 weeks the uterine body is more globular. Temperature rises ($<37.8^{\circ}\text{C}$).

Headaches, palpitations, and fainting are all commoner in pregnancy. Sweating and feeling hot are also common, due to a dilated peripheral circulation. *Management:* Increase fluid intake and take showers.

Urinary frequency is due to pressure of the fetal head on the bladder in later pregnancy. Exclude UTI.

Abdominal pain: See p 135. *Breathlessness* is common. See p 80.

Constipation tends to occur as gut motility decreases. Adequate oral fluids and a high fibre diet help combat it. Avoid stimulant laxatives—they increase uterine activity in some women. Increased venous distensibility and pelvic congestion predispose to *haemorrhoids* (if they prolapse, rest the mother head down, apply ice packs and replace them), and *varicose veins*. Resting with feet up, and properly worn elastic stockings help.

Reflux oesophagitis and heartburn occur as pyloric sphincter relaxation allows irritant bile to reflux into the stomach. Cigarettes and spices should be avoided, small meals taken, and antacids may be used. Use more pillows, and a semi-recumbent position.

Third trimester backache: Due to pelvic ligament and muscle relaxation, pain tends to be worse at night. A firm mattress, flat shoes, standing with back straight, and pelvic support from physiotherapy all help.

Carpal tunnel syndrome (p 662) in pregnancy is due to fluid retention. Advise wrist splints until delivery cures the problem.

Itch/itchy rashes are common (up to 25%) and may be due to the usual causes (OHCM p 62, check LFTs—see p 158) or to pruritic eruption of pregnancy (PEP = *prurigo of pregnancy*)—an intensely itchy papular/plaque rash on the abdomen and limbs. PEP is most common in first pregnancies beyond 35 weeks' gestation. Emollients and weak topical steroids ease it. Delivery cures it. If vesicles are present, think of *pemphigoid gestationis* (PG): a rare (1:50,000) condition which may cause fatal heat loss and cardiac failure; the baby may be briefly affected; refer early (prednisolone may be needed).¹ PG may recur in later pregnancies.

Ankle oedema: This is very common, almost normal manifestation of pregnancy. Measure BP and check urine for protein (pre-eclampsia, p 96). Check legs for DVT. It often responds to rest and leg elevation. Reassure that it is harmless (unless pre-eclampsia).

Leg cramps 33% get cramp, the latter half of pregnancy, severe in 5% often worse at night. Raising the foot of the bed by 20cm will help.

Chloasma: This is a patch of darker pigmentation, eg on the face: p 578

Nausea affects ~80%. *Vomiting* occurs in ~50%. It may start by 4 weeks and declines over the next weeks. At 20 weeks 20% may still vomit. Most respond to frequent small meals, reassurance, and a stress-free environment. It is associated with good outcome (fewer fetal losses).

Hyperemesis gravidarum

This is rare (1/1000). Risk is increased in youth, non-smokers, primips, working outside home, multiple, or molar pregnancy (hence the idea that excessively high hCG levels may be the cause—whereas steeply rising oestrogens may cause the very common phenomenon of morning sickness).¹

Presentation: Inability to keep food or fluids down; weight↓ (2–5kg) ± nutritional deficiency, dehydration, hypovolaemia, electrolyte disturbance with hyponatraemic shock, polyneuritis (B vitamins↓), behaviour disorders, liver and renal failure.

Tests: Do pcv and u&e to help guide iv fluid regimen. TFTs (there may be associated thyroid or adrenal dysfunction).² Chart losses. Exclude UTI. Ultrasound scan to exclude twins or hydatidiform mole.

Treatment: Admit to hospital. Spend time optimizing psychological wellbeing. Is she worried about how her other children are being cared for?

Most settle with due *care and attention*. If not, parenteral anti-emetics may be tried (eg metoclopramide 10mg/8h iv—but there are problems with extrapyramidal effects). Placing a Dobhoff nasogastric tube for enteral feeding has proponents,³ but note that iso-osmolar feeds are recommended, as is regular aspiration of the stomach (there is likely to be delayed gastric emptying); monitor plasma glucose (expect some insulin resistance).¹ If an iv infusion is used give iv thiamine to prevent Wernicke's encephalopathy (see *OHCM* p 736) developing.⁴ Beware rapid reversal of hyponatraemia which can cause fatal central pontine myelinosis.² Those resistant to conventional treatments may respond to steroid treatment.²

Parenteral nutrition may be needed—*OHCM* p 456. ► Get a dietitian's help.

1 C Ven 1997 *Lancet* **349** 445 [09040568] 2 M Swiet 1994 *Br J Obs Gynae* **101** 1013 [07999712]
3 J Hsu 1996 *Obstet Gynecol* **88** 343 [08752236]
4 DoH 1996 *Report on Confidential Enquiry into Maternal Deaths in the UK 1991–93* HMSO

Booking criteria and home delivery

Most women in the UK have 'shared obstetric care'—ie most of their antenatal care is from the GP and community midwife, with limited (or no) visits (usually 2) to the hospital to see the consultant under whose care they are delivered, returning home (eg after 6–72h) for postnatal care. A minority receive their complete care from hospitals and the usual reasons for this would be that their GP did not provide obstetric services, or that the complications of pregnancy made full consultant care more desirable. Some women are cared for by community midwifery staff and their GP, but increasingly, low-risk women are receiving all their care from midwives, with medical staff involvement only if complications arise. Delivery may be at hospital, in GP units (which may or may not be geographically isolated from consultant units), or, rarely, at home. ► There is quite good evidence that consultant input into antenatal care of normal pregnancies achieves no added benefits (p 95)—but risk factors making specialist visits and booking desirable are generally agreed. They divide into those relating to the mother, to her previous deliveries, and to her pregnancy now, eg:

Features pertaining to the mother

- >35yrs old (or nullip <20 or 35yrs)
- History of infertility
- From the 5th pregnancy onwards
- Height <154cm (158cm primips)
- Very obese; social deprivation
- +ve serology (HBsAg, HIV) p 98

Features of this pregnancy

- Cardiac or thyroid disease
- Multiple pregnancy
- Rh-ve with antibodies (p 242)

Features pertaining to previous deliveries

- Preterm or small (<37 weeks, <2.5kg)
- Deformity, stillbirth, neonatal death
- Caesarean section or hysterotomy
- Retained placenta; abruption
- Births requiring pelvic floor repair
- Instrumental deliveries; PPH
- Poor fetal growth or wellbeing
- Diabetes, ↑BP, anaemia
- Malpresentations after 34 weeks
- Raised serum α-fetoprotein

Is it safe for low-risk mothers to deliver in high-technology hospitals? Here interventions with their complications are more common. In the UK this question is usually academic as most GPs are reluctant to conduct births—and 6 months' Senior House Officer training in obstetrics gives scant skill. The rising birth rate and service pressures are putting these big hospitals under great strain. In places (eg New Zealand, Holland) where delivery outside of big hospitals is the norm, there is fairly clear (but not uncontested*) evidence that on all measures, and in all but the highest risk groups, big hospitals come out less favourably. The few trials seeming to favour high-technology are now recognized to be seriously flawed.

Home delivery This is rare in the UK (~1% of births, of these only 2/3 are booked in advance for home delivery). We cannot draw statistical conclusions from the sparse data comparing morbidity and mortality in home vs hospital delivery, as home deliveries are so rare. But an important observation is that rapid intervention is necessary to save life (maternal or fetal) in ~5% of low-risk pregnancies. This pinpoints the need for any domiciliary service to have good equipment available for home deliveries and good emergency back-up (provided in future by emergency obstetric ambulance units—specially trained ambulance personnel who, it is to be hoped, will liaise directly with senior medical obstetric staff in hospital).

Birthroom birth This offers a homely birth in congenial surroundings, with labour ward facilities nearby, if needed. The mother is attended by her GP and community midwife. A randomized trial has shown that mothers' satisfaction is great, and nearly all requested this type of delivery for future births. This may offer a compromise to adherents of home delivery.

Issues surrounding home delivery

- *One must remember that normal delivery is a retrospective diagnosis.*
- In the UK, because of past hospital experience of many abnormal labours, GPs are often very wary of home birth.
- Medico-legal aspects tend to dominate thoughts about worse-case scenarios—so that few GPs willingly take on intrapartum care.
- It's not clear who is to do the doctor's ordinary work when absent on a home delivery—if a mother particularly wants their GP present. Some small surgeries have had to close during delivery as no locum was available—an unacceptable consequence of offering mothers extra choice. NB: in the UK, midwives, but not GPs, have a statutory duty to help at home deliveries.
- It is not clear if there is a ready supply of doctors or midwives with the necessary experience in suturing and neonatal resuscitation. Where there is a good team, there is no doubt that home delivery can be a safe and rewarding experience.
- It has to be remembered that decisions about the place of labour are dynamic, and need revising (eg in 29%) as events in pregnancy unfold. It is not always clear how these decisions are to be taken, and how they are to be communicated.
- Necessary equipment is not readily available, eg Entonox[®],—and we have not been able to get blood on site so as to be prepared for possible exsanguination.
- The key factor in increasing choice about home delivery is a good working relationship between the parents, the GP, and the midwife. Where this exists, ~70% of home delivery requests tend to come to fruition; where the GP is rated as being unsupportive, in a UK context, this figure drops to 54%.
- Everyone needs to know that transfer in labour is common in labours starting off as planned home deliveries (14%)—but that this need not mean that there is excess morbidity.
- It is salutary to note that in 1994–5 only 18.5% of deliveries in England and Wales were considered 'normal' and without antenatal or postnatal complications.

medline:9518893

Pre-pregnancy counselling

The aim is to help prospective parents embark upon pregnancy under conditions most likely to ensure optimal wellbeing for the fetus. Babies conceived 18–23 months after a live birth have the lowest rate of perinatal problems.¹ Ensure that a woman is rubella (and chickenpox, p 216) immune prior to pregnancy. Other areas covered include:

- Optimal control of chronic disease (eg diabetes) before conception.
- Discontinuation of teratogens prior to conception (p 161).
- Medication to protect the fetus from abnormality (eg folate supplement for neural tube defects, p 228 and below).
- Provision of expert information for those at known risk of abnormality so pregnancy or its avoidance is an informed choice and any tests needed (eg chorionic villus sampling, p 166) are planned. Regional genetic services give detailed pre-pregnancy counselling. See p 210–2. In relevant ethnic populations, take blood for thalassaemia and sickle-cell tests (p 154).
- If past/family history of thromboembolism, screen for thrombophilia.²

For all women advice concerning the following may be relevant:

Diet To prevent neural tube defects and cleft lip all should have folate-rich foods and folic acid 0.4mg daily (eg Preconceive®) from before conception—until 13 weeks' gestation (5mg/day po if history of neural tube defect, some epileptic drugs p 161). These foods have >0.1mg of folic acid per serving: Brussels sprouts, asparagus, spinach, blackeye beans, fortified breakfast cereals. Avoid liver and vitamin A (vitamin A embryopathy risk).

94 Cigarette smoking may decrease ovulations, result in abnormal sperm production, reduce sperm penetrating capacity, ↑rates of miscarriage (2-fold), and is associated with preterm labour and lighter-for-dates babies (mean is 3376g in non-smoker; smoker: 3200g). Reduced reading ability in smokers' children up to 11yrs shows that longer term effects may be important. About 17% of smoking mothers stop before or in pregnancy.

Alcohol consumption High levels of consumption are known to cause the fetal alcohol syndrome (p 226). Moderate drinking has not been shown to adversely affect the fetus but alcohol does cross the placenta and may affect the fetal brain. Miscarriage rates are higher among drinkers of alcohol. How to cut consumption: see p 454.

Spontaneous abortion (SA) At least 12% of first pregnancies spontaneously abort. Rates after 1 SA are increased to ~24%; after 2 SA to ~36%; after 3 to ~32% and after 4 to ~25%, so chances of a future pregnancy succeeding are ~2 in 3. Pregnancy order of SA/live pregnancies is also relevant: the more recent a live birth the more likely next time will be successful.

Recurrent spontaneous abortion/miscarriage See p 23.

Pre-pregnancy counselling is best done by those who will look after the pregnant woman. Those with chronic medical conditions may require liaison between obstetrician and physician. Computerized practices can identify beneficiaries of pre-pregnancy counselling for these conditions:^{1,2}

- | | | | |
|-----------------------|------------------------------|--------|---------------------|
| • Diabetes mellitus | • Epilepsy | • BP ↑ | Genetic history eg: |
| • Tropical travellers | • Rubella-susceptible | • SLE | • Spina bifida etc |
| • Frequent abortion | • Pet-owners (toxoplasmosis) | • PKU | • Thalassaemia |
| | | | • Duchenne's |
| | | | • Cystic fibrosis |

Antenatal care

The aims of antenatal care are to: •Detect any disease in the mother •Ameliorate the discomforts of pregnancy •Monitor and promote fetal wellbeing •Prepare mothers for birth •Monitor trends to prevent or detect early any complications of pregnancy: BP is the most important variable (eclampsia, p 96).

Who should give antenatal care? Not all women need see a doctor at least once per pregnancy or labour: midwives do very well on their own (with fewer interventions), calling in doctors *only if a specific need arises*.

The 1st antenatal visit is very comprehensive. ►Find a language interpreter if she needs one. Avoid using relatives (confidentiality issues). *History:*

- Usual cycle length; LMP (a normal period?); Naegele's rule: expected delivery date \approx 1yr and 7 days after LMP minus 3 months—(not if last period a pill withdrawal bleed; for cycles shorter than 28 days subtract the difference from 28; if longer, add the difference from 28). A revised rule suggests the addition of 10 days rather than 7 is more accurate.¹
- Contraception; drugs; past history, eg surgery to abdomen or pelvis.
- Any fertility problems; outcome and complications of past pregnancies.
- Is there family history of diabetes, BP \uparrow , fetal abnormality, or twins?
- If past or family history of DVT or embolism, screen for thrombophilia.²
- Is she poor (eg gas/electricity supply cut off)? *Unmarried? Unsupported? Subject to domestic violence?* (See p 455.) *A substance abuser?* (See p 362.)
- Is the diet nutritious? Advise on avoiding pâtés and soft cheese (*OHCM* p 593) and vitamin overdose, eg no vitamin tablets or liver (p 94).

Examination: Check heart, lungs, breasts, nipples, BP, weight, and abdomen. Cervical smear is needed. Any varicose veins?

Tests: Blood: Hb, group (antibodies if Rh-ve, p 242), syphilis and rubella serology, sickle test if black, α -fetoprotein/triple test (p 166 & p 228), Hb electrophoresis + HBsAg (p 100 & p 154) if relevant. Offer HIV test, with counselling (*OHCM* p 584). MSU for protein, glucose, and bacteria. If she is foreign or is a TB contact or is a hospital worker, consider CXR after 14 weeks.

Ultrasound: in many units an ultrasound scan is routine (and an event for the parents) at 18 weeks, to confirm dates, look for fetal abnormalities, and exclude ovarian cysts. If the mother is over 35 years old, consider amniocentesis at \sim 17 weeks or other tests to exclude Down's syndrome (p 166).

Suggest: Parentcraft/relaxation classes; dental visit. Enquire about problems and anxieties. Consider need for *iron and folate* (p 139 and p 154).

Advise on: Smoking, alcohol, diet, correct use of seat belts, and taking adequate rest. Ensure knowledge of social security benefits. Usual exercise and travel are ok (avoid malarious areas) up to 34 weeks on most airlines (see thromboprophylaxis p 81). Intercourse is fine if no vaginal bleeding.

Later visits Check weight, urine for albumin and glucose, BP, fundal height, fetal movements (primips feel them \sim 19 weeks, earlier if multip), fetal heart—and lie and presentation from 32 weeks. Do Hb, eg at 28 & 36 weeks; look for Rh antibodies. Visits are monthly from 16 to 32 weeks, every 2 weeks until 36 weeks; then weekly. There is evidence that this is too often for most tests, but not often enough for BP and albuminuria.

The head is usually engaged (p 84) by 37 weeks in caucasian primips (if not, consider: large (or malpositioned) head, small pelvis or obstruction, placenta praevia, or wrong estimation of dates). At 36 weeks some check the pelvis clinically in primips. For favourable pelvis see page 86.

Hypertension in pregnancy

Pre-existing essential hypertension Drugs may need modifying (some ↓ fetal growth). medline:10675164 Risk of pre-eclampsia is raised $\times 5$ (suspect if BP rises by 30/15mmHg from baseline or hyperuricaemia, proteinuria, or clotting activation). *If symptoms are episodic, think of pheochromocytoma.*

Pre-eclampsia Terminology: see p 76. This is pregnancy-induced hypertension (PIH) with proteinuria \pm oedema. It is a multisystem disorder originating in the placenta. The primary defect is failure of trophoblastic invasion of spiral arteries, so the arterial wall does not distend enough to allow sufficient blood flow to the placenta in late pregnancy, and increasing BP is a mechanism which partially compensates for this. Pre-eclampsia also affects hepatic, renal and coagulation systems. It develops after 20 weeks and usually resolves within 10 days of delivery. Eclampsia (1 in 2000 maternities) is a major cause of maternal death and fetal morbidity/mortality. Pre-eclampsia may be asymptomatic, so frequent screening is vital. It may recur in a subsequent pregnancy.

Risk factors: Maternal: Past or family history of pre-eclampsia; sexual union for <4 months (or primips, or IVF by ICSI, p 57—all ?from no sperm-mediated partner-specific immune tolerance 2002 medline:11879865); <155cm tall; weight ↑; age <20 or >35; past migraine; BP ↑; renal disease. Incidence is lower in smokers.

Fetal: Hydatidiform mole (↑BP at 20 weeks); multiple pregnancy; placental hydrops (eg rhesus disease). Fetal causes all have increased placental bulk.

Effects of pre-eclampsia: Plasma volume ↓; peripheral resistance ↑; placental ischaemia. If the BP is >180/140mmHg microaneurysms develop in arteries. DIC may develop. Oedema may develop suddenly (eg weight ↑ by 1kg suddenly). Proteinuria is a late sign, meaning renal involvement, which may be detected earlier by doing urate levels (>0.29mmol/L at 28 weeks >0.34mmol/L at 32 weeks, and >0.39mmol/L at 36 weeks suggest pre-eclampsia). Initially glomerular filtration is normal and only serum urate is ↑ later urea and creatinine increase. The liver may be involved (contributing to DIC)—and HELLP syndrome may be present (p 158) with placental infarcts. Fetal asphyxia, abruption, and small babies (p 104 & p 119) may also occur.

Preventing eclampsia Antenatal BP checks/urinalysis; use of MgSO₄ if risk ↑. Uterine artery Doppler scans can identify high-risk women in whom aspirin use results in a significant reduction in pre-eclampsia. meta-analysis medline:11704184

Symptomatic pre-eclampsia may mimic 'flu, with headache, chest or epigastric pain, vomiting, and pulse ↑—but also visual disturbance, shaking hyperreflexia, and irritability. The mother is now in danger of generalized seizures (eclampsia) and treatment must occur. Death may be imminent from stroke (commonest), hepatic, renal, or cardiac failure. ▶ Prophylactic magnesium sulfate halves risk of eclampsia, and may ↓ maternal deaths there don't seem to be substantive short term side-effects (mother or baby). Maggie study: N=10,141 Lancet 2002 187

Management *Early phase:* Admit if BP rises by >30/20mmHg over booking BP; if BP \geq 160/100; if \geq 140/90 with proteinuria, or there is growth restriction. In hospital, measure BP 2–4-hourly, weigh daily, test all urine for protein, monitor fluid balance, check U&E, LFTs, urate & placental function, and platelets (beware falls to $<110 \times 10^9/L$) regularly. Also do regular antenatal cardiotocography (p 116) with us scan to check growth and biophysical profiles. In asymptomatic pre-eclampsia, ↑BP may be treated with drugs, eg methyldopa: 250mg—1g/6h under supervision in hospital if BP rises to 170/110mmHg, in order to buy time for fetal maturation, and if all other variables are satisfactory. If signs worsen deliver the baby (liaise with paediatricians). Delivery is the only cure. Give all pre-eclamptic women H₂-blockers at onset of labour

►► Managing severe pre-eclampsia

This applies to those with BP >160/110 with proteinuria >0.5g/day or BP ≥140/90 with proteinuria plus one or more of:

- Seizures
- Headache or epigastric pain
- Platelets <100 × 10⁹/L
- Visual disturbance
- Creatinine >100μmol/L
- ALT >50iu/L
- Clonus (>3 beats)
- Creatinine clearance <80mL/min.

- Continuously monitor maternal oxygen saturation, and BP every 15mins.
- Use 'Maggie' régime of prophylactic magnesium sulfate: 4g (8mL of a 50% solution) ivi over 15min in 100mL 0.9% saline; then maintenance as below.
- Catheterize for urine output; hourly T^o; FBC, U&E, LFTs, creatinine every 12h.
- Monitor fetal heart rate; assess liquor volume and fetal growth by scan; umbilical cord doppler if possible.
- *Delivery is the only cure for these women.* When a decision is made to deliver, contact on-call consultant, anaesthetist, and senior labour ward midwife.

Treatment of hypertension

- If BP >160/110mmHg or mean arterial pressure (MAP) >125mmHg, use hydralazine 5mg iv slowly unless pulse >120bpm, repeating every 20mins until 20mg total given; if needed. Alternative: labetalol 20mg iv increasing after 10min intervals to 40mg then 80mg until 200mg total is given.
- Give prophylactic H₂ blockers until normal postnatal care starts.
- Restrict fluids to 85mL/h. Watch urine output hourly. If oliguria <100mL/4h consider giving 500mL human albumin solution (HAS) ivi. Consider HAS also prior to hydralazine use, caesarean section, or use of regional anaesthesia. If HAS is used, consider inserting central venous pressure (cvp) line: if further oliguria, one is recommended. If cvp >10mmHg, give 20mg furosemide (or if there is persistent oliguria). If central venous pressure is <10mmHg consider giving 500mL HAS by ivi.

Treatment of seizures (eclampsia):

- Treat a first seizure with 4g magnesium sulfate in 100mL 0.9% saline ivi over 5min + maintenance ivi of 1g/h for 24h.
- If there are recurrent seizures give 2g ivi magnesium sulfate as a bolus.
- Check tendon reflexes and respiratory rate every 15min.
- Stop magnesium sulfate ivi if respiratory rate <14/min or SaO₂ <95%.
- Monitor Mg²⁺ levels (therapeutic range: 2–4mmol/L) if recurrent fits or renal compromise; have iv calcium gluconate ready in case of MgSO₄ toxicity.
- Use diazepam if fits continue at consultant's discretion (eg 5–10mg slowly iv). If seizures continue consider other causes (consider CT scan).

Pitfalls in the management of eclampsia and pre-eclampsia

- Belief that the disease behaves predictably, and that BP is a good marker.
- Ignoring mild proteinuria; even if 1+, the patient may be dead within 24h.
- Believing that antihypertensives stop pre-eclampsia. They don't. Delivery is the only cure. Diuretics may further deplete the already reduced plasma volume, and are especially contraindicated (except in the rare left ventricular failure or laryngeal oedema complicating pre-eclampsia).
- Believing that delivery removes risk. In the UK, half of eclamptic fits are postpartum: half of these are >48h postpartum. Continue vigilance until clinically and biochemically normal. Avoid early discharge.
- Ergometrine should not be used for the third stage (oxytocin may be used). Ergometrine further increases BP so would risk stroke.
- Not replacing significant blood loss *meticulously*. Risks are hypovolaemia or fatal fluid over-load ± acute respiratory distress syndrome. Have one person (the most experienced) in charge of all the ivs.
- Failure to inform anaesthetists early and use intensive care facilities.

Antenatal infection*

Rubella Childhood vaccination prevents rubella-susceptibility. ►Asymptomatic reinfection can occur making serology essential in all pregnant rubella contacts. Routine antenatal screening finds those needing puerperal vaccination (avoid pregnancy for 3 months: vaccine is live). Symptoms (p 214) are absent in 50%. The fetus is most at risk in the first 16 weeks' gestation. 50–60% of fetuses are affected if maternal primary infection is in the first month of gestation: <5% are affected if infection is at 16 weeks. Risk of fetal damage is much lower (<5%) with re-infection. Cataract is associated with infection at 8–9 weeks, deafness at 5–7 weeks (can occur with second trimester infection), cardiac lesions at 5–10 weeks. Other features: purpura, jaundice, hepatosplenomegaly, thrombocytopenia, cerebral palsy, microcephaly, mental retardation, cerebral calcification, microphthalmia, retinitis, growth disorder. Abortion or stillbirth may occur. If suspected in the mother seek expert help. Compare serum antibody levels 10 days apart and look for IgM antibody 4–5 weeks from incubation period or date of contact.

Cytomegalovirus (cmv) In the UK, CMV is a commoner cause of congenital retardation than rubella. Maternal infection is usually mild or asymptomatic, or there is fever ± lymphadenopathy and sometimes sore throat. Up to 5/1000 live births are infected, of whom 5% will develop early multiple handicaps, and have cytomegalic inclusion disease (with non-specific features resembling rubella syndrome, plus microcephaly, choroidoretinitis). Another 5% later develop cerebral calcification (IQ↓); sensorineural deafness and psychomotor retardation. Diagnosis (paired sera) may be difficult. Amniocentesis at >20 weeks can detect fetal transmission. medline:10645866 Also test throat swabs, urine culture, and baby's serum after birth. Reducing women's exposure to toddlers' urine (the source of much infection) during pregnancy can limit spread. NB: reactivation of old CMV may occur in pregnancy; it rarely affects the baby. One way to know that +ve serology does not reflect old infection is to do serology (or freeze a sample) pre-pregnancy.

Toxoplasmosis 40% of fetuses are affected if the mother has the illness (2–7/1000 pregnancies); the earlier in pregnancy the more the damage. Symptoms are similar to glandular fever. Fever, rash, and eosinophilia also occur. If symptomatic, the CNS prognosis is poor. Diagnose by reference laboratory IgG and IgM tests. *Maternal Rx (Royal College Regimen)*: Start spiramycin promptly in infected mothers, eg 1.5g/12h po. In symptomatic non-immune women test every 10 weeks through pregnancy. In those infected consider fetal cord blood sampling, eg at 21 weeks for IgM to see if the fetus is infected. If the fetus is infected, give the mother pyrimethamine 50mg/12h as loading doses on day 1, then 1mg/kg/day + sulfadiazine 50mg/kg/12h + calcium folinate 15mg twice weekly all until delivery. *Affected babies*: (diagnose by serology.) Oral pyrimethamine 2mg/kg/day for 2 days, then 1mg/kg/day + sulfadiazine 50mg/kg/12h + calcium folinate 15mg twice-weekly. Give this for six 4-week courses, each separated by 4 weeks of spiramycin 50mg/kg/12h. Prednisolone is given until signs of CNS inflammation or choroidoretinitis abate (eg 0.5mg/kg/12h). Prevention includes avoiding eating raw meat, washing hands after touching raw meat, wearing gloves when gardening or dealing with cat litter, and avoiding sheep during lambing time.

HIV Babies have a 15% chance of acquiring HIV if the mother is +ve (higher risk in Africa; but HIV-2 is less likely than HIV-1 to be passed on). Offer mothers tests for HIV, (see counselling *OHCM* p 584). Serology depends on detecting IgG and IgM antibodies (false +ve in up to 1.6%). Those with +ve serology should **not** be given steroids to accelerate fetal lung maturity.

Vertical transmission occurs during vaginal delivery (1st twins are twice as commonly infected as 2nd), and caesarean section protects, eg halving neonatal infection, as does zidovudine 300mg + lamivudine 150mg twice daily from 36 weeks, with a stat dose of each at the onset of labour, then zidovudine 300mg/3h + lamivudine 150mg/12h until delivery—then reverting to the prenatal dose for 7d, while giving the baby zidovudine 4mg/kg + lamivudine 2mg/kg/12h. This works well only when combined with other methods of reducing mother-to-baby spread. Bottle feeding ↓vertical transmission (?by up to 50%, but is unwise if hygiene is a problem). ⁻⁰⁰²medline:11955535 &: 10459952

Serology is difficult as transferred maternal antibodies persist up to 18 months in uninfected infants—but gene amplification via a polymerase chain reaction can detect neonatal HIV proviral sequences. Culture is also possible—but not all infected children give +ve cultures at birth, and specialized investigations may need to be performed, eg the detection of IgA (which does not cross the placenta) by modified Western blotting.

Intrauterine HIV is associated with prematurity and growth restriction (in developing countries, but not in Europe). Clinical illness appears sooner than in adult AIDS (eg at aged 6 months)—with hepatosplenomegaly, failure to thrive, encephalopathy, recurrent fevers, respiratory diseases (interstitial lymphocytic pneumonitis), lymphadenopathy, salmonella septicaemia, pneumocystis, and CMV infection. Death is usually from respiratory failure or overwhelming infection. Mortality: 20% at 18 months. ▶ For treatment and advice on how to prevent transmission in labour, see p 155.

Intrauterine syphilis Routine maternal screening occurs (in the UK, numbers needed to screen to prevent 1 case = 55,700—in some parts of London 2:1000 women are infected); if active infection is found treat the mother with procaine penicillin 600mg/24h IM daily for 10 days. Neonatal signs: rhinitis, snuffles, rash, hepatosplenomegaly, lymphadenopathy, anaemia, jaundice, ascites, hydrops, nephrosis, meningitis, ± keratitis, and nerve deafness. Nasal discharge exam: spirochaetes; x-rays: perichondritis; CSF: ↑Monocytes and protein with +ve serology. *Treatment:* Give procaine penicillin 37mg/kg/24h IM for 3 weeks.

Listeria This affects 6–15/100,000 pregnancies—more during epidemics. Maternal symptoms: fever, shivering, myalgia, headache, sore throat, cough, vomiting, diarrhoea, vaginitis. Abortion (can be recurrent), premature labour, and stillbirth may occur. Infection is usually via infected food (eg milk, soft cheeses, pâté). ▶ Do blood cultures in any pregnant patient with unexplained fever for ≥48h. Serology, vaginal and rectal swabs do not help (it may be commensal there). See *OHCM* p 593.

Perinatal infection usually occurs in the second or third trimester. 20% of affected fetuses are stillborn. Fetal distress in labour is common. An early postnatal feature is respiratory distress from pneumonia. There may be convulsions, hepatosplenomegaly, pustular or petechial rashes, conjunctivitis, fever, leucopenia. Meningitis presents more commonly after perinatal infection. Diagnosis is from culture of blood, CSF, meconium, and placenta. Infant mortality: 30%. Isolate baby (nosocomial spread can occur). Treat with ampicillin 50mg/kg/6h IV and gentamicin 3mg/kg/12h IV until 1 week after fever subsides. Monitor gentamicin levels.

Sheep-borne conditions Listeriosis, toxoplasmosis, and ovine chlamydia (*Chlamydia psittaci*) can be contracted from sheep. Ovine chlamydial infection is rare; can cause DIC, septicaemia, and renal failure in pregnant women and abortion of the fetus. Diagnose by serology; treat with erythromycin or tetracycline. ▶ Pregnant women should not handle sheep or lambs.

Previously called 'TORCH' infections: toxoplasmosis, other (eg syphilis), rubella, CMV, herpes (and hepatitis). The 1st 4 are acquired antenatally—herpes & hepatitis usually perinatally. Other agents are important (chickenpox; HIV, p 216–7). See also Erythrovirus (= Parvovirus, p 214)—zoster reactivation in pregnancy is not a risk for the baby.

Perinatal infection

Hepatitis B virus (HBV) Although chronic carriers were uncommon in the UK, drug addiction and immigration make it commoner (0.3–17/1000 deliveries). Some now recommend that all mothers be screened. If the mother develops acute infection in the mid- or third trimester there is high risk of perinatal infection. Her risk of death is 0.5–3%. Most neonatal infections occur at birth. Most infected neonates will develop chronic infection and in infected males lifetime risk of developing hepatocellular cancer is 50%; 20% for ♀. Most will develop cirrhosis—so immunization is really important. ► Give infants of known carriers hepatitis immune globulin 0.5mL IM within 12h of birth, and hepatitis B vaccine 0.5mL (eg Engerix B® 10µg at a different site) within 7 days and at 1 and 6 months. medline:10426262 & 10072416

Hepatitis E Risk of maternal mortality is ↑; death is usually postpartum, preceded by coma and massive PPH. A vaccine is awaited. medline:8160022 & 8404882

Herpes hominis 80% is due to herpes simplex type II virus. 50% of babies will be infected during birth if the mother has active cervical lesions. Take viral cultures weekly (from 36 weeks) from the cervix of all women with a history of herpes. If virus is present, aim for elective caesarean section. When membranes rupture spontaneously still perform caesarean if within 4h of rupture. If a mother has active lesions culture baby's nasopharyngeal secretions at 24–48h. If positive treat with aciclovir. Neonatal infection usually appears at 5–21 days with vesicular pustular lesions, often at the presenting part or sites of minor trauma (eg scalp electrode). Periocular and conjunctival lesions may occur. With systemic infection encephalitis (focal fits or neurological signs), jaundice, hepatosplenomegaly, collapse, and DIC may occur. Infected neonates should be isolated and treated with aciclovir (p 256). Seek expert help.

Chlamydia trachomatis Associations: low birth weight, premature membrane rupture, fetal death. ~1/3 of infected mothers have affected babies. Conjunctivitis develops 5–14 days after birth and may show minimal inflammation or purulent discharge. The cornea is not usually involved.

Complications: Chlamydia pneumoniae, pharyngitis, or otitis media.

Tests: Immunofluorescence or culture. Giemsa smear: many inclusions.

Treatment: 1% tetracycline ointment or drops 6-hourly for 3 weeks, + erythromycin 10mg/kg/6h po for ~3 weeks eliminates lung organisms. Give parents/partners erythromycin. medline:11095596

Gonococcal conjunctivitis Occurs within ~4 days of birth, with purulent discharge and lid swelling, ± corneal hazing, corneal rupture, and panophthalmitis. **Treatment:** Infants born to those with known gonorrhoea should have cefotaxime 100mg/kg IM stat, and chloramphenicol 0.5% eye-drops within 1h of birth. For active gonococcal infection give benzylpenicillin 50mg/kg/12h IM and 3-hourly 0.5% chloramphenicol drops for 7 days. Isolate the baby.

Ophthalmia neonatorum This is purulent discharge from the eye of a neonate <21 days old. Originally referring to *Neisseria gonorrhoea* there are many causes: chlamydiae, herpes virus, staphylococci, streptococci, pneumococci, *E. coli*, and other Gram -ve organisms. **Tests:** In a baby with a sticky eye take swabs for bacterial and viral culture, microscopy (look for intracellular gonococci) and chlamydia (eg immunofluorescence). Treat gonococcus and chlamydia as above; other infections with neomycin drop or ointment (allows chlamydia detection—not so with chloramphenicol).

Clostridium perfringens Suspect this in any complication of criminal abortion—and whenever intracellular encapsulated Gram +ve rods are seen on genital swabs. It may also complicate death *in utero*, and it may also infect

haematomas, and other anaerobic sites. *Clinical features:* Endometritis → septicaemia/gangrene → myoglobinuria → renal failure → death.

Management: • Surgically debride all devitalized tissue. • Hyperbaric O₂.

• High-dose iv benzylpenicillin (erythromycin if serious penicillin allergy).

The use of gas gangrene antitoxin is controversial. Seek expert help.

Tuberculosis All babies born into families with a member who has TB, to immigrant mothers from communities with a high incidence of TB, or who will be travelling to areas with a high incidence of TB should be given BCG (Bacillus Calmette–Guérin) vaccination after birth 0.05mL intradermally at deltoid insertion: if using a multiple puncture gun dose is 0.03 mL (percutaneous). Usual other childhood vaccinations should be given at the normal time but avoid using the BCG vaccinated arm for 3 months. Babies born to mothers with active or open TB should be separated from the mother until she has had 2 weeks of treatment and is sputum negative. BCG vaccinate the baby and treat with isoniazid until it has a positive skin reaction. Mothers with inactive or healed TB should have a CXR in pregnancy to exclude reactivation.

For perinatal **listeria** and **streptococcal infection**, see p 260.

Prematurity

► This is a leading cause of perinatal mortality and morbidity.

Premature infants are those born before 37 weeks' gestation. Prevalence: ~6% singletons, 46% of twin, 79% of triplet or higher order deliveries. About 2% are before 32 weeks—when neonatal problems are greatest. In 25%, delivery is elective (p 110). 10% are due to multiple pregnancy; 25% are due to APH, cervical incompetence, amnionitis, uterine abnormalities, diabetes, polyhydramnios, pyelonephritis, or other infections. In 40% the cause is unknown, but abnormal genital tract colonization (bacterial vaginosis) with ureaplasma and *Mycoplasma hominis* is implicated, either as a risk factor or risk marker. We also know that consumption of fish oil in pregnancy can ↑ birth weight by ↓ risk of recurrence of preterm delivery.*

Management of preterm rupture of membranes Admit; take MSU, and HVS—using a sterile bivalve speculum. If liquor is not obvious its presence is suggested if nitrazine sticks (which are pH-sensitive) turn black (false +ve with infected vaginal discharge, semen, blood, and urine). In 80% membrane rupture will be followed by onset of labour. The problem with the 20% who do not go into labour is balancing the advantages of remaining *in utero* for increasing maturity and surfactant production against the threat of infection (which accounts for 20% of neonatal deaths after premature rupture of membranes). Intrauterine infection supervenes after membranes have ruptured in 10% within 48h, 26% by 72h, 40% by >72h. Prophylactic antibiotics may allow labour to be delayed. If infection develops, take blood cultures and give iv antibiotics (eg ampicillin 500mg/6h iv + netilmicin 150mg/12h iv) and expedite labour (p 110). Antibiotics within 24h before labour, reduce rates of intraventricular haemorrhage and periventricular malacia (below) in the baby. If labour supervenes it should be allowed to progress. In the few cases where liquor ceases to drain for more than 48h the mother can be gradually mobilized. medline:10901551

Management of preterm labour In 50% contractions cease spontaneously. Treating the cause (eg pyelonephritis) may make it cease. Attempts to suppress contractions (tocolysis) are unlikely to succeed if membranes are ruptured or the cervix >4cm dilated. Use between 24–33 weeks. Consider transfer to hospital with scbu facilities. Call paediatrician to attend to the baby at birth. See cord-cutting recommendations p 82.

Tocolytic drugs Absolute CI: chorioamnionitis, fetal death or lethal abnormality, condition (fetal or maternal) needing immediate delivery. Relative CI: fetal growth restriction or distress, pre-eclampsia, placenta praevia, abruptio, cervix >4cm. **Ritodrine regimen:** Simpler than salbutamol, but not always available—see OPPOSITE. Continue infusion for 12–48h after contractions cease. Monitor: maternal pulse, BP, T°, contractions, fetal heart. Salbutamol is an alternative (se similar). Nifedipine 30mg po stat (+20mg po 90min later if still contracting), then 20mg/8h po is as effective. SE: ↓ BP, headache, flushing, transient tachycardia; myocardial infarction is reported. medline:991722

Glucocorticoids Betamethasone or dexamethasone 12mg/12h IM twice promotes fetal surfactant production, lowering mortality and complications of RDS (p 240) by 40–50%; they also help close patent ductuses and protect against periventricular malacia, a cause of cerebral palsy. Use before 34 weeks only. Avoid if severe pre-eclampsia, infection, or if HIV+ve. Benefit is maximal after 24h; effects last a week.

*S Olsen 2002 *BMJ* i 447 [011859044] If fish oil intake is low, small amounts of n-3 fatty acids provided as fish or fish oil may protect against prematurity & low birth weight

Prematurity, survival, and disability—the figures

- One major, landmark question is: *Is the baby over 28 weeks' gestation?*
- The disability rate is 25% if gestation is <28 weeks—but half this if gestation is 28–29 weeks).
- 10% of those who survive at gestations <28 weeks will never be independently mobile, or communicate intelligibly with others.
- Only 4% of babies born before 24 weeks will survive, and of survivors, >50% will be severely disabled.
- Use of surfactant (p 240) has not reduced the viability threshold, or rates of severe disability in babies born at <28 weeks' gestation.
- Of babies born >30 weeks' gestation, in the absence of deformity, ~100% now survive.
- Ritodrine may protect against grade III–IV periventricular haemorrhage in infants 24–32 weeks' gestation, <1.5kg birthweight. ²⁰⁰¹medline:11420315

Guidance for infusion rates of ritodrine

Syringe pump (Add 3×5mL ampoules of ritodrine to 35mL of 5% dextrose.)	Controlled infusion device (Add 3×5mL ampoules ritodrine to 500mL of 5% dextrose.)
---------------------------------------------------------------------------	---------------------------------------------------------------------------------------

Dose	Rate	Dose	Rate
50 micrograms/min	1mL/hour	50 micrograms/min	10mL/hour
100 micrograms/min	2mL/hour	100 micrograms/min	20mL/hour
150 micrograms/min	3mL/hour	150 micrograms/min	30mL/hour
200 micrograms/min	4mL/hour	200 micrograms/min	40mL/hour
250 micrograms/min	5mL/hour	250 micrograms/min	50mL/hour
300 micrograms/min	6mL/hour	300 micrograms/min	60mL/hour
350* micrograms/min	7mL/hour	*350 micrograms/min	70mL/hour

*maximum recommended dose

Start with 50 micrograms/min. Increase by 50 micrograms/min every 15min until contractions cease or side-effects (maternal pulse >140 beats/min, FHR↑ by >20 beats/min, maternal systolic BP falls by >20mmHg, tremor, nausea, vomiting, flushing). Usual dose: 150–350 micrograms/min. Total daily intravenous dose should not exceed 120 milligrams. Continue infusion for 12–48h after contractions cease.

Problems with tocolytics The main problems are hyperglycaemia and pulmonary oedema from fluid overload. Use only on specialist advice in those with known heart disease. Avoid in those prone to supraventricular tachycardia, eg with Wolff–Parkinson–White or Lown–Ganong–Levine syndromes. Atosiban appears as effective as β -agonists but with fewer side-effects. ²⁰⁰¹medline:11236112

Monitoring

- Maternal pulse and blood pressure every 15min.
- Maternal temperature and blood glucose 4-hourly (eg BM-Test® sticks).
- Monitor contractions and fetal heart rate.
- Keep strict fluid records (input/output charts).
- Check U&E every 24h (to avoid hypokalaemia) or more often if indicated. Check ECG if hypokalaemic.
- Auscultate lung fields regularly. Monitor maternal O₂ saturation if equipment is available.
- Chest pain, dyspnoea, or dry cough should lead to immediate cessation of infusion.

Small for gestational age (SGA)

►When talking to parents, avoid the term *growth retardation* as this may imply to them the inevitability of mental handicap—which is not the case. Distinguish premature babies from those who are small-for-gestational age (SGA): they are at risk from different problems after birth.

Causes of growth restriction (IUGR) Growth restricted neonates are those weighing < the 10th centile for their gestational age (*Tables*, p 294).

Maternal factors: Multiple pregnancy, malformation, infection, smoking, diabetes, BP ↑, Hb ↓, pre-eclampsia, heart or renal disease, asthma. medline:1067516
Those having intensive shared-care with an expert have less hypoxia, and less intra-uterine growth restriction (IUGR). medline:10217076

~10% are to mothers who only ever produce small babies.

Asymmetric growth restriction: Where placental insufficiency was the cause, head circumference is relatively spared (the baby has been starved).

Antenatal diagnosis 50% are undetected before birth, and many babies suspected of IUGR do not have it. Measuring fundal height progress from the symphysis pubis is a reasonable method of measuring growth, especially if used with centile charts. Oligohydramnios (p 120) and poor fetal movements are other indications of placental insufficiency. If growth restriction is suspected, growth *in utero* can be monitored by serial ultrasounds of head circumference and abdominal circumference. If umbilical cord Doppler blood flows are normal the outcome of growth restricted pregnancies is better (fewer premature births and stillbirths). Those with abnormal Dopplers may benefit from maternal low-dose aspirin (p 119), eg 75mg/24h po. Biophysical profile monitoring (p 118) and antenatal cardiotocography (p 116) are used to try to detect those babies who are becoming hypoxic *in utero* and who would benefit from delivery. Advise the mother to stop smoking, to use a fetal kick chart (p 116), and to take plenty of rest.

Labour and aftercare Growth restricted fetuses are more susceptible to hypoxia, so monitor in labour (p 116). After birth, temperature regulation may be a problem, so ensure a warm welcome; nurse those <2kg in an incubator. After being relatively hypoxic *in utero* the Hb at birth is high, so jaundice is more common. They have little stored glycogen so are prone to hypoglycaemia. Feed within 2h of birth and measure blood glucose before each 3-hourly feed. If hypoglycaemic despite regular feeds, transfer to a special care unit. They are more susceptible to infection. Birth reveals those for whom abnormality was the cause of growth restriction.

Distinguishing growth restriction from prematurity Before 34 weeks' gestation there is no breast bud tissue, from then it develops at 1mm diameter/week. Ear cartilage develops between 35 and 39 weeks so premature babies' ears do not spring back when folded. Testes lie in the inguinal canal at 35 weeks, in the scrotum from 37. Labia minora are exposed in premature girls. Skin creases on the anterior 1/3 of the foot appear by 35 weeks (on anterior 2/3 by 39, and all over from 39). 'Premis' have red, hairy skin. Vernix is made from 28 weeks and is maximal at 36 weeks. Premis do not lie with legs flexed until 32 weeks. All limbs are flexed from 36 weeks.

Effects of IUGR in adult life Many studies have confirmed ↑ risk from:

- Hypertension
- Coronary artery disease
- Non-insulin-dependent diabetes mellitus
- Autoimmune thyroid disease

So fetal malnutrition casts a long shadow. Specific early deficiencies are important too (eg iodine, iron).

Large for gestational age

These are babies above the 90th centile in weight for gestation.

Causes: Constitutionally large (usually familial—the largest 10% of the population); maternal diabetes (p 156); hyperinsulinism; Beckwith–Wiedemann syndrome (p 742).

Labour and aftercare: Large babies are at risk of birth injury (see impacted shoulder p 128). Large babies are prone to immaturity of suckling and swallowing and may need temporary tube feeding. They are prone to hypoglycaemia and hypocalcaemia. Polycythaemia may result in jaundice. They are also prone to left colon syndrome: a self-limiting condition clinically mimicking Hirschsprung's disease (p 218) whereby temporary bowel obstruction (possibly also with meconium plug) occurs. Rarely, there is risk of renal vein thrombosis.

►► Fetal distress

Fetal distress signifies hypoxia. Prolonged or repeated hypoxia causes fetal acidosis. An early sign may be the passage of meconium in labour (p 128). Other signs that the fetus may be hypoxic are a fetal tachycardia persisting above 160 beats per minute (tachycardia may also occur if the mother has a high temperature or is dehydrated). Hypoxia may also be reflected by loss of variability of the baseline in the fetal heart rate trace and slowing and irregularity of the heart rate (especially late decelerations—p 116). ►► If the heart rate falls below 100 beats per minute urgent assessment is required. Hypoxia may be confirmed by the use of fetal blood sampling (p 116). When significant hypoxia appears to be present (eg pH <7.24), deliver promptly (by the quickest route available, eg caesarean section or vaginal extraction). In complete anoxia the pH falls by 0.1 unit per minute.

►► Obstetric shock

Most obstetric shock is associated with severe haemorrhage. It should be remembered that with placental abruption, actual bleeding may be far in excess of that revealed *per vaginam* (p 108). Other causes of shock may be: ruptured uterus (p 134), inverted uterus (p 140); amniotic fluid embolus (p 143), pulmonary embolism, adrenal haemorrhage, and septicaemia.

Septicaemia may lack classical signs (eg pyrexia) and must be considered where profound persisting shock is present, be appropriately investigated (eg blood cultures), and treated, eg amoxicillin/clavulanic acid 1g/6h iv + metronidazole 500mg/8h ivi (+ gentamicin 1.5mg/kg/8h iv given over >3min; do levels, but not needed acutely, see *OHCM* p 549). Prompt resuscitation is required (see individual pages for management). Renal function and urine output should always be measured after shock has occurred (p 160). A late complication can be Sheehan's syndrome (also called Simmonds' disease) whereby pituitary necrosis leads to lack of thyroid-stimulating hormone, adrenocorticotrophic hormone, and the gonadotrophic hormones, hence leading to hypothyroidism, Addisonian symptoms and genital atrophy.

Postmaturity (prolonged pregnancy)

Prolonged pregnancy is defined as that exceeding 42 completed weeks or more from the LMP. Affecting 5–10% of pregnancies it is associated with increased perinatal mortality (5/1000 if 37–42 weeks, 9.7/1000 if >42 weeks). The dangers are placental insufficiency, and problems during labour due to the fetus being larger (25% are >4000g), the fetal skull more ossified and less easily moulded, the passage of meconium more common (25–42% labours), and fetal distress more common. Stillbirth rate is $\uparrow \times 4$, neonatal deaths $\uparrow \times 3$, and neonatal seizures $\uparrow \times 10$ in pregnancies going beyond 42 weeks compared with birth between 37 and 42 weeks.

Antepartum monitoring with use of kick charts (p 116), cardiotocography, and biophysical profiles (p 118) can be used to try to detect fetuses who may be becoming hypoxic. Doppler studies of cord blood flow may be used to look for absent end diastolic flow as a predictor of fetal compromise. Without evidence to suggest problems, induction should be recommended from 41+ weeks after which time there are higher rates of caesarean section for those managed 'conservatively'. Use ultrasound dates not LMP for calculating gestation. 'Membrane sweeps' remain a valid non-pharmacological prelude to induction; more women go into spontaneous labour if as much membrane as possible is swept from the lower segment at examination between 41–42 weeks than if examined unswept. 6 women will be membrane swept for 1 formal induction avoided. meta analysis 1999
medline:10430199

Induction with vaginal prostaglandins may be associated with lower rates of instrumental delivery than induction with oxytocin. It is estimated that 500 inductions are needed to prevent 1 perinatal death. Vaginal misoprostol eg 50µg/4h to a maximum of 5 doses appears to be a good induction agent, although not yet routinely used as there are worries about uterine hyperstimulation. After induction, monitor the fetus in labour (p 116).

Postnatal signs of postmaturity: Dry, cracked, peeling, loose skin; decreased subcutaneous tissue; meconium staining of nails and cord.

Prelabour rupture of membranes at term

2–4% of term births are complicated by prelabour rupture of membranes without labour spontaneously starting within 24h. It is usual practice when labour does not ensue within a few hours of membrane rupture to start labour with an oxytocin ivi to reduce the possibility of ascending infection. It is helpful to give vaginal prostaglandins when the cervix appears unfavourable. Routine use of prostaglandins does not reduce the rate of caesarean section (but does reduce time to delivery). Waiting 12h after the membranes rupture before instituting oxytocin is associated with lower rates of caesarean section (rate ~11%) as opposed to early use of oxytocin (rate ~15–20%). Oral misoprostol, eg 100µg po, repeated twice, 4 hourly if needed, is as effective as oxytocin for inducing labour and reduces time to delivery in nulliparous women. 2000
medline:10688506

When delay is intended maternal temperature must be monitored and oxytocin started should pyrexia develop.

►► Antepartum haemorrhage (APH)

Traditionally this has been defined as bleeding at >24 (formerly 28) weeks' gestation. Any bleeding in pregnancy is associated with increased perinatal mortality. Severe bleeds can cause maternal death. medline:10078578

► Avoid vaginal examination: placenta praevia may bleed catastrophically.

Dangerous causes Abruptio, placenta praevia, vasa praevia (here the baby may bleed to death).

Other uterine sources: Circumvallate placenta, placental sinuses.

Lower genital tract sources: Cervical polyps, erosions and carcinoma, cervicitis, vaginitis, vulval varicosities.

Placental abruption ('accidental haemorrhage') Part of the placenta becomes detached from the uterus. The outcome depends on the amount of blood loss and degree of separation. The cause is unknown but it is associated with pre-eclampsia, may recur in a subsequent pregnancy (6%), is commoner in smokers, and may complicate external cephalic version. Bleeding may be well localized to one placental area and there may be delay before bleeding is revealed.

Consequences: Placental insufficiency may cause fetal anoxia or death. Compression of uterine muscles by blood causes tenderness, and may prevent good contraction at all stages of labour, so beware a PPH (which occurs in ~25%). Posterior abruptions may present with backache. There may be uterine hypercontractility (>7 contractions per 15min). Thromboplastin release may cause DIC (10%). Concealed loss may cause maternal shock after which beware renal failure and Sheehan's syndrome (p 106).

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Placenta praevia (For terminology and complications see p 78). The placenta lies in the lower uterine segment. Bleeding is always revealed.

Distinguishing *abruption* —————► from *placenta praevia*:

Shock out of keeping with the visible loss	Shock in proportion to the visible loss
Pain constant	No pain
Tender, tense uterus	Uterus not tender
Normal lie and presentation	Both may be abnormal
Fetal heart: absent/distressed	Fetal heart usually normal
Coagulation problems	Coagulation problems rare
Beware pre-eclampsia, DIC, anuria	Small bleeds before large

Note: the risk of PPH is increased in both conditions. The lower segment may not contract well after a placenta praevia.

Management of APH Always admit. ►► If bleeding is severe call emergency ambulance, put up ivi, take bloods, and raise legs. Give O₂ at 15L/min via mask with reservoir. On admission, if shocked give fresh ABO Rh compatible or O Rh-ve blood (eg 6u, 2 ivis) fast until systolic BP >100mmHg. Send blood for clotting screen. Catheterize bladder; keep urine output >30mL/h. Call anaesthetist to monitor fluids (cvp lines help). *Summon expert help urgently.* If bleeding is severe, *deliver*—caesarean section for placenta praevia (sometimes for abruption, or induction). Beware PPH.

For milder bleeding, set up ivi, do Hb, crossmatch, coagulation studies, and U&E. Check pulse, BP, and loss regularly. Establish diagnosis (ultrasound of placenta, speculum examination). If placenta praevia is the diagnosis, keep in hospital until delivery (usually by caesarean section at 37–38 weeks). If pain and bleeding from a small abruption settles and the fetus is not compromised the woman may go home (after anti-D, if indicated), but treat as 'high-risk' pregnancy thereafter. Arrange follow-up.

Using anti-D immunoglobulin

Dose 250u for gestations <20 weeks, 500u if >20 weeks. Give into deltoid (buttock absorption is too slow) as soon as possible after incident, at latest within 72h. After 20 weeks do Kleihauer test (FBC bottle of maternal blood; fetal RBCs therein are less susceptible to lysis, enabling them to be counted, so measuring the bleed's volume).

Postnatal use 500u is the normal dose after 20 weeks' gestation. 37% of Rh-ve women give birth to Rh-ve babies and these women do not need anti-D.

- Anti-D should be given to all Rh-ve women where the baby's group cannot be determined (eg macerated stillbirths), or if circumstances are such that the baby's group is unknown 72h post delivery.
- Do a Kleihauer test on all eligible for anti-D. 500u anti-D can suppress immunization by up to 4mL of fetal red cells (8mL of fetal blood), but 1% of women have transplacental haemorrhage (TPH) of >4mL, especially after manual removal of placenta, and with caesarean section. A Kleihauer test is especially important in stillbirth, as massive spontaneous transplacental haemorrhage can be the cause of fetal death. Where >4mL TPH is suggested by the Kleihauer screen, a formal estimation of the TPH volume is required and 500u anti-D given for every 4mL fetal cells transfused (maximum 5000u anti-D at 2 IM sites/24h). Note: Kleihauer tests can be negative where there is ABO incompatibility as fetal cells are rapidly cleared from the maternal circulation. Liaise with the Transfusion Service. Check maternal blood every 48h to determine clearance of cells and need for continuing anti-D.
- Don't give prenatal anti-D if antibodies to anti-D are present.
- Any mother receiving anti-D prenatally (see below), should also receive it postnatally unless she delivers an Rh-negative baby.

Use of anti-D in abortion in rhesus negative mothers

- 1 Anti-D should be given to all having surgical or medical terminations of pregnancy unless they are already known to have anti-D antibodies. Give 250u if <20 weeks; 500u (and Kleihauer) if > 20 weeks' gestation.
- 2 Anti-D should always be given where spontaneous abortion is followed by instrumentation.
- 3 Anti-D should be given where spontaneous complete abortion occurs after 12 weeks' gestation.
- 4 With threatened abortion after 12 weeks give anti-D; if bleeding continues intermittently give anti-D 6-weekly until delivery.
- 5 Routine anti-D is not recommended with threatened miscarriage before 12 weeks' gestation (but consider if viable fetus, heavy or repeated bleeding, and abdominal pain).

Use of anti-D in pregnancy in Rh-ve mothers

- 1 Give anti-D 500u at 28 and 34 weeks to rhesus negative women (primip antenatal sensitization falls from 1.5% to 0.2%). Anti-D may still be detectable in maternal blood at delivery. Still give postnatal anti-D, if indicated (as above).
- 2 When significant TPH may occur: with chorionic villus sampling; external cephalic version; APH; uterine procedures (eg amniocentesis); abdominal trauma. Use 250u before 20 weeks' gestation, 500u (and do Kleihauer) after 20 weeks.
- 3 Anti-D should be given in cases of ectopic pregnancy.
- 4 For threatened abortion see above.

Induction of labour

5–20% of UK labours are induced artificially, usually because it has been decided that to remain *in utero* is relatively more risky for the fetus than to be born, but in some it is because of risk to the mother. 75% of inductions are for hypertension, pre-eclampsia, prolonged pregnancy or rhesus disease. Other indications are diabetes, previous stillbirth, abruption, fetal death *in utero*, and placental insufficiency.

► Inducing mothers at 41+ weeks reduces stillbirth rates.

Contraindications Cephalopelvic disproportion which is absolute, malpresentations other than breech or face presentation, fetal distress, placenta praevia, cord presentation, vasa praevia, pelvic tumour, previous repair to cervix. Cone biopsy requires caution.

Cervical ripeness When an induction is being planned the state of the cervix will be assessed. In 95% of women at term the cervix is ripe. If primips are induced with an unripe cervix (Bishop's score ≤ 3 , see below) the rates of prolonged labour, fetal distress, and caesarean section are increased. This is less marked in multips.

Modified Bishop score	0	1	2
Cervical dilatation (cm)	0	1–2	3–4
Length of cervix (cm)	>2	1–2	<1
Station of head (cm above ischial spines)	–3	–2	–1
Cervical consistency	firm	medium	soft
Position of cervix	posterior	middle	anterior

A score of >5 is 'ripe'. An unripe cervix may be ripened using prostaglandin (PGE₂) vaginal gel (1mg) the evening before or the morning of induction (use 2mg for unfavourable primip cervix). If antenatal fetal heart rate monitoring is indicated, this should commence before prostaglandin insertion. If there is failure to ripen (occurs in 12%) PGE₂ may be repeated 6–8h later. If the cervix still remains unripe consider caesarean section. PGE₂ may stimulate uterine contractions or precipitate labour.

Once the cervix is ripe, rupture the membranes (amniotomy) and start intrapartum fetal heart rate monitoring using a scalp clip or pulse oximetry (less invasive, see OHCM p 154). Oxytocin is given iv in 5% dextrose using a pump system (eg Ivac®). Infusions start at 2 milliunits (mu) per min, doubling every 30min until effective uterine contractions are produced (usually at a rate of 4–12mu/min; occasionally 32mu/min may be necessary). Beware uterine hyperstimulation and the use of large volumes of iv fluid (if >4 litres, there is risk of water intoxication—ie confusion, convulsions, and coma). When the cervix is 5cm dilated the uterus is more sensitive to oxytocin and 8mu/min may be sufficient to maintain contractions. Note: the Dublin regimen (p 112) results in most women going into spontaneous labour. Misoprostol (a prostaglandin E₁ analogue) po or pv is as effective at cervical ripening and inducing labour as PGE₂ and oxytocin. Vaginal route shortens time to delivery but possibly results in more caesarean sections.¹

Problems of induction • Iatrogenic prematurity. • Infection (use antibiotic cover (p 152) in women with heart lesions as risk of endocarditis). • Bleeding (vasa praevia). • Cord prolapse (eg with a high head at amniotomy). • Some will lead to caesarean section.

1 J Kwon 2001 *BJOG* 108 23 [11212999]—167 women were randomized to receive misoprostol 50µg/6h po or pv until the cervix was favourable for amniotomy, spontaneous rupture of membranes, or active labour occurred.

The active management of labour

Dublin is a centre of excellence for labouring mothers because of its coherent and very successful labour management plan, which leads to a low caesarean section rate (eg 5% vs 14% for other centres). This depends on only ever having women who are actually in labour on labour ward, and regularly assessing them to check that cervical dilatation is progressing well—and taking action if it is not. Primiparous women with single, cephalically presenting babies, who are not achieving cervical dilatation satisfactorily have augmentation of uterine contractions with the use of oxytocin. Women are kept informed of their progress and are given an estimated time of delivery and are assured that delivery should be within 12h of entering the labour ward. Caesarean section is performed on those for whom delivery is not imminent at 12h. By use of this regimen it is possible to keep a caesarean section rate below 5% and a forceps rate of ~10%, with the vast majority of women effecting delivery themselves.

Labour This is defined as painful uterine contractions of increasing strength, accompanied by a 'show' or cervical dilatation. Women arriving who may not be in labour are accommodated overnight on antenatal wards. Those in labour are admitted to the labour ward and thereby committed to delivery.

Augmentation This recognizes that a primiparous uterus is an inefficient organ of birth that is *not* prone to rupture—but the multiparous uterus is an efficient organ but one which is rupture-prone. Oxytocin may be used to enhance efficiency in the primiparous uterus without maternal danger. Oxytocin may be dangerous to multips as delay is likely to be from obstructed labour, so a decision to use it must only be made by senior obstetricians.

Examination Women are examined at admission and hourly for the first 3h by a single examiner (may be by rectal examination). Progress is assessed at 1h; artificial rupture of membranes is performed to aid progress and allow liquor viewing. Examination 1h later will result in augmentation of labour by oxytocin in primips if 1cm extra cervical dilatation has not been achieved. Examination is subsequently every 2h. Oxytocin will be used if primip dilatation does not progress at 1cm/h or if there is delay in descent in the involuntary pushing phase of second stage.

Oxytocin A uterus in labour is sensitive to oxytocin. Use standard dilutions (see *BNF*), and a maximum of 5 units oxytocin in 24h.

Induction by artificial rupture of membranes (ARM). Women return to the antenatal ward to await onset of labour. Those 10% who do not go into labour spontaneously are returned to labour ward after 24h and oxytocin is used exactly as above. 90% of these women also deliver within 12h, thereby keeping caesarean section rates low.

The partogram This graphs labour's progress (p 111). Dublin uses a simple partogram with time plotted against cervical dilatation, and records fetal heart rate, state of liquor, analgesia, and oxytocin use.

Meconium in liquor At ARM this suggests placental insufficiency. Meconium other than light staining in good liquor volume prompts fetal blood sampling and scalp clip electronic monitoring—or prompt caesarean section (fetal blood pH low, or very thick meconium).

The opposite page depicts Dublin practice for primiparous mothers with term babies of vertex presentation with a normal head. Its authors stress the benefit of their policies both to mothers and also from the financial management point of view as it allows them to have a personal midwife for each mother throughout the time that she is in labour, but with unit costs per delivery of only one-third of costs of UK hospitals.

Midwife monitoring is normally every 15min using a Pinard scope; electronic monitoring is used if there is meconium staining of liquor. Membranes are ruptured so that the liquor can be regularly inspected.

Other UK units have different policies. Some argue that artificially rupturing membranes may not be entirely beneficial as the liquor cannot cushion the fetal head and there is evidence that artificial rupture of membranes is associated with more fetal heart rate decelerations. ARM may also facilitate spread of HIV from mother to baby (p 155). A review of current knowledge suggests that of early amniotomy, early use of oxytocin, and the provision of professional support throughout first labours, it is the last that is the most important for reducing rates of operative delivery. *Support* may be the key, as shown by shorter labours, fewer forceps and caesarean section deliveries, and reduced analgesia requirements of women supported by doulas (trained female companions) during labour—an effect not seen when the expectant father is the companion.

medline:10643833

One large randomized USA trial found active management of labour had no benefits, and did not reduce rates of caesarean section.

medline:7643880

Whether to institute or abandon active management is being overtaken by women's views: many reject intervention, even if it is offered.

Pain relief in labour

► Adequate pain relief in labour is the obstetrician's greatest gift to womankind. Not everyone wants natural birth—and do not make people feel guilty about requesting pain relief. A good anaesthetic agent in labour must be harmless to mother and baby, allow maintenance of maternal cooperation, and must not affect uterine contractility.

Education That given by the National Childbirth Trust meets all these criteria. Education about labour reduces fear; breathing exercises and relaxation techniques teach the mother ways to combat pain herself.

In the first stage, **narcotic injections** are often tried, eg pethidine 50–150mg IM (not if birth expected in <2–3h as neonatal respiratory depression may occur, reversible with naloxone 0.1–0.2mg as a single dose IM). Expect analgesia by 20min and to last 3h. NB: doses in the lower range frequently produce vomiting, but no relief of pain. Other SE: disorientation, ↓gastric emptying, neonatal respiratory depression. CI: mother on MAOIs (p 366). In Scandinavia **sterile water injections** are used for 1st stage low back pain. Four 0.1–0.25mL injections of sterile water are given intracutaneously adjacent to Michaelis' rhomboid (in the lumbosacral area). Initially causing pain, the injections produce a wide papule with red circumference. Pain reduction by 50% has been reported lasting 90min. Pain of injection can be reduced by use of nitrous oxide simultaneously¹ or by giving them subcutaneously.²

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Nitrous oxide (50% in O₂ = Entonox®) can be inhaled throughout labour and is self-administered using a demand valve. CI: pneumothorax.

Pudendal block (sacral nerve roots 2, 3, and 4) uses 8–10mL of 1% lidocaine (=lignocaine) injected 1cm beyond a point just below and medial to the ischial spine on each side. It is used with perineal infiltration for instrumental delivery, but analgesia is insufficient for rotational forceps.

Spinal block These may be used for rotational delivery or caesarean section. For details and contraindications, see p 776.

Epidural anaesthesia See p 777. Pain relief is by anaesthetizing pain fibres carried by T11–S5. Epidurals must only be set up once labour is established (cervix >3cm). Set up ivi first, give 500mL Hartmann's solution to prevent ↓BP. Check pulse, BP, respirations, contractions, and fetal heart every 15min after the epidural is set up. Top-ups are required ~2-hourly. Epidurals may be very helpful for the following: OP position (p 126), breech, multiple pregnancy, preterm delivery, pre-eclampsia, forceps delivery, inco-ordinate uterine contractions.

Problems: For those due to technique, see p 777. There may be postural hypotension (put up ivi, nurse 15° to left side), urinary retention (catheterize regularly), paralysis (pelvic floor muscle paralysis reduces rotation and voluntary effort in second stage, so increasing need for forceps). After delivery: urinary retention and headache (esp. after dural puncture).

Epidural anaesthesia for those receiving heparin thromboprophylaxis:³

► Be sure you understand what type of heparin the patient is using. With low dose subcutaneous heparin, wait 4h after heparin given before siting the block or removing catheter. After giving block allow at least 2h before giving next dose of heparin. Where *low molecular weight heparin LMWH* is used, wait 10–12h after heparin dose before inserting block or removing catheter; wait at least 6h after block siting before next dose of LMWH. Aim to give LMWH at 18.00h each day. Beware use of other anticoagulants or nonsteroidal anti-inflammatory drugs. All patients must be extremely

vigilantly monitored to detect new numbness; weakness; bowel or bladder dysfunction. Any neurological problem must be investigated as an emergency.

Combined spinal epidural (cse) anaesthesia gives quicker pain relief, with little or no motor blockade in most mothers, allowing standing, walking, sitting, and voiding urine. Using the intrathecal route (27G Whiteace needle) apparently does not cause a significant rise in the incidence of headache (only ~0.13%). The patient can control the dose, and this leads to a dose reduction of 35%, and reduces motor blockade. Women should inform their midwife if they notice light-headedness, nausea, or weak legs. Spontaneous delivery rates are not better than with traditional epidural. ► Skilled anaesthetic help is vital.

Transcutaneous electrical nerve stimulation (TENS) Electrodes placed on the back, give pulses stimulating large nerve fibres to the brain, aiming to block pain impulses. It needs careful instruction but is under the mother's control. In 1 meta-analysis, 6 out of 9 trials showed no effect; those that did, showed only weak effects, eg ↑time before epidural use.

Fetal monitoring

In high-risk pregnancy, antepartum cardiotocography and biophysical profiles by ultrasound (p 118) are used to monitor fetal activity and responsiveness. The aim is to detect intrauterine hypoxia prenatally.

Kick charts: In the last 10 weeks of pregnancy the mother can count the fetal movements daily and record the time at which the baby has made 10 movements. Although a poor predictor of fetal outcome, less than 10 movements in 12h may be a warning sign of problems and the mother should attend her obstetric unit for cardiotocography.

Cardiotocography: Doppler ultrasound detects fetal heart beats and a tocodynamometer over the uterine fundus records any contractions. A continuous trace is printed over ~30min (eg a paper speed of 1 or 2cm/min) with the mother lying semi-recumbent, or in the left lateral position or in the half-sitting position. A normal trace in an afebrile, mother at term who is not having drugs has a **base rate** of 110–150 beats/min, with a **variability** of 5–25 beats/min, and at least 2 **accelerations** (a common response to movement or noise) of an **amplitude** ≥ 15 beats/min over a 15–20min period. (Fetal heart rate falls by ~ 1 beat/min/week from 28 weeks.) An abnormal fetal heart rate is characterized by a baseline tachycardia of > 170 beats/min or bradycardia (< 100 beats/min), or a sinusoidal pattern. Loss of variability (< 5 beats/min over a 40min period) and recurrent late or atypical decelerations are associated with fetal compromise and are the most important prognostic indicators.

Tests need to be done every 24h antenatally to identify the changing fetal heart rate pattern associated with hypoxia.

Intrapartum monitoring Death and disability due to complications of labour occur in $< 1:300$ labours. Intrapartum fetal heart rate monitoring aims to detect patterns known to be associated with fetal distress. The diagnosis of fetal distress may be supported by the passage of fresh meconium and fetal hypoxia on blood sampling.

The fetal heart rate is traditionally monitored by using a Pinard stethoscope between contractions, but with this method decelerations related to contractions may be missed.

Continuous fetal heart rate monitoring has poor predictive value, over-diagnosing fetal distress even if used with fetal blood sampling. Its value is uncertain even in high-risk labours—but it is widely used. Meta-analyses suggest that routine monitoring \downarrow rates of neonatal seizures, but increases operative deliveries.¹ Monitoring policies vary. Low-risk labours may be monitored intermittently. If high-risk, monitor throughout labour, ideally with scalp electrode or pulse oximetry. **Indications:** Abnormal admission trace; antenatal problems; use of oxytocin; fresh meconium (p 128).

Disadvantages: Limited maternal mobility and discomfort.

Management of a poor trace:

- 1 Lie the mother on her left side and give O_2 . Stop oxytocin.
- 2 Take fetal blood sample. If you do not have this facility, consider rapid delivery if the trace does not improve.

Fetal blood sampling: Fetal acidosis reflects hypoxia. Scalp blood pH of 7.3–7.4 is normal. If 7.25–7.29 repeat after 45min. If 7.2–7.24 consider caesarean section. Levels < 7.2 require immediate delivery unless in second stage when a level as low as 7.15 may be acceptable.

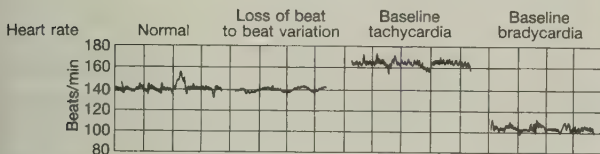
Fetal heart rate patterns and their clinical significance

The normal pattern is described opposite. Accelerations suggest intact sympathetic activity and are rarely associated with hypoxia.

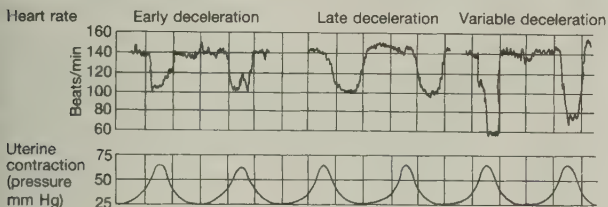
Loss of baseline variability Baseline variability of >5 beats/min shows response to vagal tone, sympathetic stimuli, and catecholamines in a well oxygenated fetal brainstem. Loss of baseline variability may reflect a preterm fetus who is sleeping, drug effects (eg diazepam, morphine, phenothiazine), or hypoxia.

Baseline tachycardia Heart rate >170 beats/min is associated with maternal fever, or β -sympathomimetic drug use, chorioamnionitis (loss of variation too), and acute/subacute hypoxia. Persistent rates >200 are associated with fetal cardiac arrhythmia.

Baseline bradycardia A heart rate <110 beats/min is rarely associated with fetal hypoxia (except in placental abruption). It may reflect \uparrow fetal vagal tone, fetal heart block, or, if spasmodic, cord compression.



Early decelerations coinciding with uterine contractions reflect increased vagal tone as fetal intracranial pressure rises with the contraction. **Late decelerations**, when the nadir of the deceleration develops some 30sec after the peak of the uterine contraction, reflect fetal hypoxia, the degree and duration reflecting its severity. **Variable decelerations**, both in degree and relation to uterine contractions, may represent umbilical cord compression around the limbs or presenting part.



Ultrasound

In each ultrasound examination, examine placental site and structure, assess liquor volume and fetal structure, and wellbeing (see below).

Early in pregnancy If there is bleeding and pain, ultrasound can confirm intrauterine pregnancy—at 4 weeks + 3 days for regular 28 day cycle with transvaginal scan, (or ectopic) and the viability of the fetus. Where there is discrepancy between uterine size and dates, gestation can be estimated, a viable fetus ascertained (not missed abortion, p 22), or twins (and if mono-chorionic or dichorionic see p 122) diagnosed—especially important after the use of fertility drugs or in the presence of hyperemesis gravidarum (also to exclude hydatidiform mole).

Estimation of gestational age Crown–rump length is measured from 6–12 weeks (~10mm at 7 weeks; ~55mm at 12 weeks). From 12 weeks the biparietal diameter can be measured (and femur length from 14 weeks so that there are 2 independent estimations with each procedure). Biparietal diameter measurements to estimate age are most accurate up to 20 weeks (unreliable from 34 weeks). Knowledge of gestational age is important in management of rhesus disease and in diabetic pregnancy (p 156)—and also helps if the date of the LMP is not known, or the cycle is irregular.

Fetal abnormality See *fetal nuchal translucency*, p 167. Many units offer routine scans to find abnormality at 18–20 weeks. With the best machines and sonographers, an increasing range of markers of fetal abnormality are discernible, eg nuchal thickness in Down's syndrome, but the sign is also +ve in ~6% of normal pregnancies. The more such markers, the greater the chance of abnormality (see p 167 for 'soft signs'). It is hard to know how to counsel women before these discerning scans: full informed consent is time-consuming, and itself causes psychopathology (p 168) and it seems odd to ask the mother what sort of informed consent she would like (but see p 169). Another problem is that many fetuses with chromosomal abnormalities suggested by early scans will spontaneously miscarry in subsequent weeks, and one will have overburdened the mother needlessly. Specific indications for scanning for abnormality include:

- Family history of neural tube defect
- Maternal diabetes or epilepsy
- AFP abnormal
- Oligohydramnios
- Twins
- Polyhydramnios

Fetal echocardiography Offered to high risk groups (personal or first degree relative history of heart disease; associated with nuchal thickness increase) it is traditionally offered at specialist centres at 22–24 weeks. Scanning at 12–15 weeks at these centres may exclude most major abnormalities.¹

Biophysical profile scoring Ultrasound of the fetus in the womb over a period of up to 30min aims to assess if the fetus is being affected by acute or chronic asphyxia. With acute asphyxia the fetus loses active biophysical variables regulated by central nervous system outflow (fetal breathing, gross body movement, fetal flexor tone, and heart rate accelerations with fetal body movement). Reduction in amniotic fluid demonstrated by pockets of fluid of less than 1cm depth measured in 2 perpendicular planes is taken to indicate chronic asphyxia. By use of specific criteria for these 5 variables, a 'wellbeing' score can be reached. Management protocols according to score then help guide the obstetrician as to when intervention should take place.

Ultrasound is used as an adjunct to the diagnostic procedures of amniocentesis, fetoscopy, cordocentesis, and chorionic villus biopsy.

In pregnancies where fetal growth is of concern (p 104) growth can be monitored by regular scans; the abdominal to skull circumference ratio being of interest. Fetal weight can be estimated when planning vaginal delivery of breech presentation (but less accurate for larger fetuses).

In later pregnancy, the lie and presentation of the fetus can be determined. If there is APH, placenta praevia can be excluded and the placenta visualized to look for abruption. With secondary postpartum haemorrhage, retained products of conception may be visualized.

Side-effects Ultrasound is one of the safest tests ever invented; nevertheless, over the years, fears have been expressed about inducing childhood cancers. There is now good evidence that this does not occur. There is one randomized trial indicating that *repeated* ultrasound with Doppler is associated with an increased risk (1.65) of birth weights below the third centile.

Doppler ultrasound and fetal wellbeing

Doppler ultrasound is a technique that can be used to assess circulation on both sides of the placenta. It has not been shown to be of use as a screening tool in routine antenatal care but it has been shown to be of use in high-risk pregnancies. Waveform outlines (velocimetry) have been seen to be abnormal in small babies who ultimately died or were severely ill perinatally, and babies known to be small from real-time ultrasound have been shown not to be at risk if umbilical artery waveforms were normal, but if end-diastolic signals are absent the baby is likely to be hypoxic and acidotic. In management of growth-retarded babies uncomplicated by other obstetric problems surveillance using Doppler velocimetry is more cost-effective than use of cardiotocography, and may be sufficient as the sole extra means of surveillance (in addition to ultrasound examination of biophysical parameters). ►Consider caesarean section if umbilical artery Doppler velocimetry shows absent or reversed end diastolic velocities in pregnancies complicated by intrauterine growth restriction and hypertension—to prevent postnatal problems such as cerebral haemorrhage, anaemia, and hypoglycaemia. These changes correlate with placental intervillous space ischaemia and spasm or occlusion of tertiary stem arterioles on the fetal side of the circulation. Placental intervillous ischaemia leads to centralization of fetal circulation so that blood returning from the placenta is shunted to fetal brain, coronary arteries and adrenals. Intervention studies based on knowledge of abnormal umbilical waveform patterns have resulted in babies small-for-gestational age (SGA) with abnormal waveform patterns being ~500g larger than expected when the mothers were given low-dose aspirin rather than placebo from the time of diagnosis. It is also the case that low-dose aspirin delays the onset of hypertension and reduces its severity in primiparous mothers whose babies had abnormal umbilical waveforms. However, aspirin has not been shown to be of benefit in low-risk pregnancies.

Amniotic fluid

Amniotic fluid (liquor) Made by amniotic cell secretion and maternal plasma filtrate, volumes are abnormal if the fetus cannot swallow it or contribute to it as urine. By 10 weeks the volume is 30mL, by 20 weeks 300mL, by 30 weeks 600mL. Maximal between 34 and 38 weeks (800–1000mL) it reduces thereafter by 150mL per week.

Oligohydramnios Liquor volume is <500mL. It is rare. medline:9448386

Associations: Prolonged pregnancy, prolonged membrane rupture, placental insufficiency, fetal urethral aplasia or renal agenesis. The (fatal) Potter's syndrome comprises fetal low-set ears, renal agenesis, hypoplastic lungs, and amnion nodosum (clumps of fetal squames).

Polyhydramnios (hydramnios) Amniotic fluid exceeds 2 litres in 1 in 250 pregnancies. 50% of cases are associated with fetal abnormality, 20% with maternal diabetes. In 30% no cause is found.

Fetal causes: Anencephaly (no swallowing reflex), spina bifida, umbilical hernia, ectopia vesicae, oesophageal or duodenal atresia, hydrops fetalis, hyper-extended attitude.

Maternal causes: Diabetes mellitus, multiple pregnancy. Presenting early in the third trimester, the mother may develop breathlessness and oedema. An abdominal girth of >100cm is suggestive. Ultrasound (p 118) is used to exclude multiple pregnancy and abnormalities. Polyhydramnios predisposes to premature labour, malpresentation, cord prolapse, placental abruption when membranes rupture, and PPH (the uterus does not contract well). As indomethacin given to the mother reduces fetal urine production it can be used to treat polyhydramnios (eg 25mg/6h po). Fetal urine production returns to normal 24h after treatment stops. During labour, check early for cord prolapse. After delivery pass a nasogastric tube in the baby to check the oesophagus is patent.

Amniocentesis Amniocentesis is the sampling of amniotic fluid. It is used for prenatal diagnosis of abnormality and in the management of rheus disease (p 242). Amniocentesis for abnormality diagnosis is carried out at 15–16 weeks' gestation, when there is enough liquor for reliable fetal cell culture and time to terminate the pregnancy in the light of results if this is desired. Early amniocentesis at 10–13 weeks can provide satisfactory samples, but has high rates of fetal loss (eg ~5%), and may be associated with talipes equinovarus. Chromosome results take 3–4 weeks to be ready. Ultrasound enables sampling from a good pool of liquor avoiding placental penetration. With sterile technique remove 15mL of fluid with a 21G needle. Give anti-D 250u IM to Rh-ve women afterwards. Counsel the woman beforehand.

Indications for amniocentesis: Advanced maternal age (≥ 35 –37yrs for Down's syndrome); a previous child with a neural tube defect (1:20 subsequently affected) or raised maternal serum α -fetoprotein (p 168 & p 228)—though ultrasound is often now used instead to look for neural tube defects; when a parent carries a balanced chromosomal translocation (1 in 4–10 chance fetus affected); risk of recessively inherited metabolic disorder (p 210); when the mother carries an X-linked disorder (to determine fetal sex). Chorionic villus sampling (p 166) can provide earlier results for many of the above, allowing earlier termination of pregnancy in the light of results. The miscarriage rate after amniocentesis later, ie 16 week amniocentesis, is ~1%. See p 166.

Dynamics of amniotic fluid and the English way of life

The English instinctively admire the intrauterine way of life. Being dark and surrounded by water appeals to their embryonic sense of humour. And the irony of being protected from alien worlds by nothing more than a circulating system of one's own urine is not lost on them. The fish is in the sea, and the sea is in the fish: even more so, the fetus is in her liquor and the liquor is in the fetus. It is this quicksilver sense of irony that shapes their sphere and keeps England afloat on seas no longer ruled. As one Englishman has said 'we are conceived in irony. We float in it from the womb. It's the amniotic fluid. It's the silver sea. It's the waters at their priestlike task washing away guilt and purpose and responsibility. Joking but not joking. Caring but not caring. Serious but not serious. . .'

Alan Bennett *The Old Country*, ISBN 0-571-19442-7, p 263

The silver sea referred to reflects Shakespeare's England as 'This precious stone set in the silver sea' nursing 'this teeming womb of royal kings' (*Richard II*, II i)

Multiple pregnancy

Incidence^{uk} Twins: 1/105 pregnancies (33% monozygotic); triplets: 1/10,000.

Predisposing factors Previous twins; family history of twins (dizygotic only); increasing maternal age (<20yrs 6.4/1000, >25yrs 16.8/1000); induced ovulation and *in vitro* fertilization; race origin (1/150 pregnancies for Japanese, 1/36 in Nigerian Igbo women). The worldwide rate for monozygotic (monochorionic) twins is constant at 3–5/1000.

Features Early pregnancy: uterus too large for dates; hyperemesis. Later there may be polyhydramnios. The signs are that >2 poles may be felt; there is a multiplicity of fetal parts; 2 fetal heart rates may be heard (reliable if heart rates differ by >10 beats/min). Ultrasound confirms diagnosis (and at 10–14 weeks, can distinguish monochorionic from dichorionic twins).¹

Complications during pregnancy Polyhydramnios (p 120); pre-eclampsia is more common (10% in singleton pregnancies; 30% in twins); anaemia is more common (iron and folate requirements are increased). There is an increased incidence of APH (6% for twins, vs 4.7% for singletons) due to both abruption and placenta praevia (large placenta).

Fetal complications Perinatal mortality for twins is 36.7/1000 (8/1000 if single; 73/1000 for triplets; and 204 for higher multiples). The main problem is prematurity. Mean gestation for twins is 37 weeks, for triplets 33 weeks. Growth restricted babies (p 104) are more common (growth rate the same as for singletons up to 24 weeks but may be slower thereafter). Malformation rates are increased 2–4 times, especially in monozygotic twins. Ultrasound is the main diagnostic test. Selective fetocide (eg with intracardiac potassium chloride) is best used before 20 weeks if indicated. With monozygotic twins, intermingling blood supply may result in disparate twin size and one being born plethoric (hence jaundiced later), the other anaemic. If one fetus dies *in utero* it may become a fetus papyraceous which may be aborted later or delivered prematurely.

Complications of labour PPH is more common (4–6% in singletons, 10% in twins). Malpresentation is common (cephalic/cephalic 40%, cephalic/breech 40%, breech/breech 10%, cephalic/transverse (Tv) 5%, breech/Tv 4%, Tv/Tv 1%). Rupturing of vasa praevia, increased rates of cord prolapse (0.6% singleton, 2.3% twins), premature separation of the placenta and cord entanglement (especially monozygous) may all present difficulties at labour. Despite modern technology some twins remain undiagnosed, staff are unprepared, and syntometrine may be used inappropriately, so delaying delivery of the second twin. Epidural anaesthesia is helpful for versions.

Management

- Ensure adequate rest (need not entail admission).
- Use ultrasound for diagnosis and monthly checks on fetal growth.
- Give additional iron and folate to the mother during pregnancy.
- More antenatal visits, eg weekly from 30 weeks (risk of eclampsia ↑).
- Tell the mother how to identify preterm labour, and what to do.
- Consider induction at 40 weeks. Have an IVI running in labour and an anaesthetist available at delivery. Paediatricians (preferably one for each baby) should be present at delivery for resuscitation should this be necessary (second twins have a higher risk of asphyxia).

Monozygous or dizygous? Monozygous twins are always same-sex and the membrane consists of 2 amnions and 1 chorion (if in doubt send for histology).

Triplets are difficult . . . more than simply difficult

Bringing up one child is difficult: bringing up twins is often very very difficult—but bringing up triplets is more than very very very difficult: it is frequently a source of significant psychopathology. During infertility consultations, eager couples may say in one breath: “We wouldn’t worry about twins. . . delighted—even triplets would be ok. . .” But how do they know, and would they find the following information chastening?

Even 4 years after their birth, all mothers in one study² suffered from exhaustion and emotional distress. The relationships with the children was often difficult (aggression and conflicts). One-third of mothers had sufficient depression to require psychotropic medication, and one-third spontaneously expressed regrets about having triplets. But do not conclude from this that it will necessarily be right to offer mothers selective termination. After triplets have been reduced to twins *in utero*, subsequently one-third of mothers will suffer emotional problems (persistent sadness and guilt) up to one year. However, adjustment had occurred in ~90% by 2 postnatal years.³

Breech (ie buttock) presentations

Breech presentation is the commonest malpresentation. Although 40% of babies are breech at 20 weeks, 20% will be at 28 weeks, and only 3% by term. It is normal in pregnancy for the buttocks to come to lie in the fundus. Conditions predisposing towards breech presentation: contracted pelvis, bicornuate uterus, fibroid uterus, placenta praevia, oligohydramnios, spina bifida, or a hydrocephalic fetus. Ultrasound may show the cause and influence the management.

Extended breech presentation is commonest—ie flexed at the hips but extended at the knees. **Flexed breeches** sit with hips and knees both flexed so that the presenting part is a mixture of buttocks, external genitalia, and feet. **Footling breeches** are the least common. The feet are the presenting part and this type has the greatest risk (5–20%) of cord prolapse.

Diagnosis of breech presentation should be made antenatally. The mother may complain of pain under the ribs. On palpation the lie is longitudinal, no head is felt in the pelvis, and in the fundus there is a smooth round mass (the head) which can be ballotted, a sensation akin to quickly sinking an apple in a bowl of water.

External cephalic version (ECV)—turning the breech by manoeuvring it through a forward, less commonly backward, somersault. Turn the baby only if vaginal delivery planned. Traditionally carried out at 34–36 weeks' gestation, it can be used from 37 weeks using tocolysis. Success rate ~53%.¹ Version contraindications: placenta praevia, multiple pregnancy, APH, small-for-dates babies, and in mothers with uterine scars, pre-eclampsia, or hypertension (the risk of abruption is increased), who have a bad obstetric history, unstable lie.¹ Give anti-D (500u) to rhesus-negative patients and take Kleihauer test (p 109) to detect the 1% who may need additional anti-D.

Mode of delivery Planned caesarean section provides better outcome for the fetus vs vaginal delivery. A large multicentre trial ($n=2088$) using experienced breech deliverers found that one baby avoided death or serious morbidity for every 14 extra caesarean sections done. Serious maternal complications were similar in both groups.²

Assisted breech delivery The breech engages in the pelvis with the bitrochanteric diameter (9.5cm) transverse. With further descent through the pelvis, rotation occurs so the bitrochanteric diameter lies antero-posteriorly as it emerges from the birth canal, being born by lateral flexion of the trunk. External rotation then occurs so that this diameter is again transverse. The shoulders enter the pelvis with the bisacromial diameter transverse, rotate through 90° emerging in the AP diameter. The head enters the pelvis with the sagittal suture transverse and rotates 90°. When the body is completely born it is allowed to hang for about 1–2 minutes until the nape of the neck is well seen. The body is then lifted above the vulva by an assistant, the head being delivered with forceps.

► Check baby for hip dislocation (↑incidence): also, if vaginal delivery, for Klumpke's paralysis (p 724) and signs of CNS injury.

Other malpresentations

Occipitoposterior presentation (OP) In 50% of patients the mothers have a long 'anthropoid' pelvis. Diagnosis may be made antenatally by palpation (p 84). On vaginal examination the posterior fontanelle will be found to lie in the posterior quadrant of the pelvis. Labour tends to be prolonged because of the degree of rotation needed, so adequate hydration and analgesia (consider epidural) are important. During labour 65% rotate 130° so that the head is occipitoanterior at the time of birth, 20% rotate to the transverse and then arrest ('deep transverse arrest'), 15% rotate so that the occiput lies truly posterior and birth is by flexion of the head from the perineum. Although in 73% delivery will be a spontaneous vaginal delivery, 22% will require forceps and 5% a caesarean section.

Face presentation Incidence 1:500. 15% are due to congenital abnormality such as anencephaly, tumour of or shortened fetal neck muscles. Most occur by chance as the head extends rather than flexes as it engages. Antenatal diagnosis: the fetal spine feels S-shaped, the uterus is ovoid without fullness in the flanks and there is a deep groove between the occiput and the back. On early vaginal examination, the nose and eyes may be felt but later this will not be possible because of oedema. Most engage in the transverse (mentobregmatic diameter $\approx 9.5\text{cm}$). 90% rotate so that the chin lies behind the symphysis (mentoanterior) and the head can be born by flexion. If the chin rotates to the sacrum (mentoposterior), caesarean section is indicated.

Brow presentation This occurs in 1:2000 deliveries and is often associated with a contracted pelvis or a very large fetus. Antenatal diagnosis: the head does not engage (mentovertical diameter $\approx 13\text{cm}$) and a sulcus may be felt between the occiput and the back. On vaginal examination the anterior fontanelle and supraorbital ridges may be felt. Deliver by caesarean section.

Transverse lie (compound shoulder presentation) This occurs in 1 in 400 deliveries and is usually in multiparous women. Other predisposing factors: multiple pregnancy; polyhydramnios; in primips: arcuate or septate uterus, placenta praevia, contracted pelvis. Antenatal diagnosis: ovoid uterus wider at the sides, the lower pole is empty, the head lies in one flank, the fetal heart is heard in variable positions. On vaginal examination with membranes intact no distinguishing features may be felt, but if ruptured and the cervix dilated, ribs, shoulder, or a prolapsed hand may be felt. The risk of cord prolapse is high. External cephalic version (p 124) may be attempted from 32 weeks. If malpresentation persists or recurs caesarean section will be necessary. Those with persistent instability of lie need hospital admission from 37 weeks (to prevent cord prolapse at home when the membranes rupture) and decision as to elective caesarean section.

►► Prolapsed cord

Cord prolapse is an emergency because of the risk of cord compression causing fetal asphyxia. There is an increased incidence at twin deliveries, footling breech delivery, and with shoulder presentations. If a cord presentation is noted prior to membrane rupture, carry out caesarean section. Always be aware when artificially rupturing membranes, when the presenting part is poorly applied, that cord prolapse is a possible risk.

Management The aim is to prevent the presenting part from occluding the cord. ►► This may be effected by:

- Displacing the presenting part by putting a hand in the vagina; push it back up (towards mother's head) during contractions.
- Using gravity, either place the woman head down or get her into knee-elbow position (kneeling so rump higher than head).
- Infuse 500mL saline into bladder through size 16 catheter.
- Keep cord in vagina: do not handle it (to prevent spasm).

If the cervix is fully dilated and the presenting part is sufficiently low in pelvis, deliver by forceps (if cephalic) or by breech extraction (by a suitably experienced obstetrician). Otherwise arrange immediate caesarean section, if the fetus is still alive. There are few other emergencies where speed is so vital.

►► Impacted shoulders (shoulder dystocia)

This is inability to deliver the shoulders after the head has been delivered.

Associations: Large/postmature fetus; malformation; short cord. medline:9746371

Management The danger is death from asphyxia ►► Speed is vital as the cord is usually squashed at the pelvic inlet. Make a large episiotomy, then:

- Use McRoberts' (hyperflexed lithotomy) position. medline:11513914 &:10554757
Abduct, rotate outwards, and flex maternal femora so each thigh touches the abdomen (1 assistant to hold each leg). This straightens the sacrum relative to the lumbar spine and rotates the symphysis superiorly helping the impacted shoulder to enter the pelvis without manipulating the fetus.
- Apply suprapubic pressure: with flat of hand laterally in the direction baby is facing and posteriorly. Apply steady traction to the fetal head towards the floor. This aims to displace the anterior shoulder allowing it to enter the pelvis. If this fails, check anterior shoulder is under the symphysis (here the diameter of the outlet is widest); if not rotate it to be so and repeat traction. If this fails, rotation by 180° (so posterior shoulder now lies anteriorly) may work, as may delivery of the posterior arm. If all else fails replacement of the fetal head by firm pressure of the hand to reverse the movements of labour and return the head to the flexed occipito-anterior position and caesarean delivery can be very successful. If the baby dies prior to delivery, cutting through both clavicles (cleidotomy) with strong scissors assists delivery.
- Check the baby for damage, eg Erb's palsy or fractured clavicle.

Meconium-stained liquor

In late pregnancy, it is normal for some babies to pass meconium (bowel contents), which stains the amniotic fluid a dull green. This is not significant. During labour, fresh meconium, which is dark green, sticky, and lumpy, may be passed. This may be a response to the stress of a normal labour, or a sign of distress, so transfer to a consultant unit (if in a GP unit) and commence continuous fetal heart rate monitoring (p 116). Aspiration of fresh meconium can cause severe pneumonitis. As the head is born, suck out the oropharynx and nose. Have a paediatrician in attendance (p 244) to suck out pharynx and trachea under direct vision using a laryngoscope.

Forceps

Forceps are designed with a cephalic curve, which fits around the fetal head, and a pelvic curve which fits the pelvis. Short-shanked (eg Wrigley's) forceps are used for 'lift out' deliveries, when the head is on the perineum; long-shanked (eg Neville Barnes) for higher deliveries, when the sagittal suture lies in the AP diameter. Kielland's forceps have a reduced pelvic curve, making them suitable for rotation (only in experienced hands).

Conditions of use The head must be engaged; the membranes ruptured the position of the head known and the presentation suitable, ie vertex or face (mentoanterior); there must not be cephalo-pelvic disproportion (moulding not excessive); the cervix must be fully dilated and the uterus contracting, and analgesia adequate (perineal infiltration for the episiotomy; pudendal blocks may be sufficient for mid-cavity forceps and ventouse deliveries but not for Kielland's). The bladder must be empty.

Indications for use Forceps may be used when there is delay in the second stage: this is frequently due to failure of maternal effort (uterine inertia or just tiredness), epidural analgesia, or malpositions of the fetal head. They may be used when there is fetal distress or a prolapsed cord, or eclampsia—all occurring only in the second stage. They are also used to prevent undue maternal effort, eg in cardiac disease, respiratory disease, pre-eclampsia. They are used for the after-coming head in breech deliveries.

Technique This should be learnt from demonstration. The following is an *aide-mémoire* for non-rotational forceps. Place the mother in lithotomy position with her bottom just over the edge of the delivery bed. Use sterilizing fluid to clean the vulva and perineum; catheterize; check the position of the head. Insert pudendal block and infiltrate the site of the episiotomy (not necessary if she has an epidural). Assemble the blades to check they fit, with the pelvic curve pointing upwards. The handle which lies in the left hand is the left blade and is inserted first (to the mother's left side) and then the right: the handles should lock easily. Traction must not be excessive (the end of bed is not for extra leverage!). Synchronize traction with contractions, guiding the head downwards initially. Do a large episiotomy when the head is at the vulva. Change the direction of traction to up and out as the head passes out of the vulva. If baby needs resuscitation, give to paediatrician. Give vitamin K (p 244). Is thromboprophylaxis needed? (p 81). **Complications of forceps:** Maternal trauma; fetal facial bruising, and VII paralysis (usually resolves).

Ventouse The ventouse, or vacuum extractor, associated with less maternal trauma than forceps is preferred worldwide, but not in the UK. It may be used in preference to rotational forceps because, as traction is applied, with the cup over the posterior fontanelle, rotation during delivery will occur. It can be used through a partially dilated cervix (primips should be almost fully dilated, multips >6cm), but should not be used if the head is above the ischial spines. A cup is applied with a suction force of 0.8kg/cm². The baby's scalp is sucked up to form a 'chignon', which resolves in 2 days. There is increased rate of fetal cephalhaematoma (p 144) and neonatal jaundice so give vitamin K (p 244). Maternal trauma still occurs in ~11%.

If ventouse, low forceps, or mid forceps are needed for 1st delivery, spontaneous rates for 2nd delivery are 91%, 88%, 82% respectively in the absence of induction or augmentation.¹

Caesarean section

Incidence 21% of UK labours;¹ (Hong Kong: >27%).² **Maternal mortality:** 0.33 per 1000. **Morbidity** is higher—from infection, ileus, thromboembolism, and delay in bonding—unless done under epidural anaesthesia. Factors contributing to this high section rate include over-diagnosis of fetal distress by cardiotocography, fear of litigation should brain damage follow difficult vaginal delivery, and more induced labours (oxytocin may promote fetal distress). Mothers' wishes are also an important factor: although dismissed by some as 'too posh to push' one must bear in mind that choice is one of the markers of good obstetric practice; the challenge is to make that choice *informed*.

Lower uterine segment incision The almost universal use of this incision makes uterine rupture (p 134) in subsequent pregnancy much less common, allows better puerperal healing, reduces infection as the wound is extraperitoneal, and has lower post-operative complication rates.

Classical caesarean section (vertical incision) Rarely used. Indications: •Very premature fetus, lower segment poorly formed •Fetus lies transverse, with ruptured membranes and liquor draining •Structural abnormality makes lower segment use impossible •Constriction ring present •Fibroids (some) •Some anterior placenta praevia when lower segment abnormally vascular •Mother dead and rapid birth desired.

Before an emergency section ►*Explain to the mother what is to happen*

- Activate the anaesthetist, theatre staff, porters, and paediatrician.
- Have the mother breathe 100% O₂ if there is fetal distress.
- Neutralize gastric contents with 20mL of 0.3 molar sodium citrate, and promote gastric emptying with metoclopramide 10mg iv. (NB: there is no time for H₂ agonists to work; ranitidine is kept for elective sections, eg 150mg po 2h before surgery.) Consider pre-operative emptying of stomach (eg if prolonged labour or opiate given). The stomach should be routinely emptied prior to extubation to minimize risk of postoperative aspiration. See Mendelson's syndrome, p 134.
- Take to theatre (awake); set up ivi. Take blood for crossmatch, eg 2u (6u and 2 ivis if previous section + anterior placenta praevia—see below).
- Catheterize the bladder. Tilt 15° to her left side on operating table.
- Use pulse oximetry peri- and post-operatively in those with dark skins (cyanosis is difficult to detect).
- Tell the paediatrician if the mother has had opiates in the last 4h.

Note: halothane should not be used for obstetric procedures because uterine muscle relaxation increases bleeding. Other anaesthetic problems include vomiting on induction (see rapid sequence induction, p 770), and light anaesthesia (out of consideration for the baby) causing paralysis/awareness. ►In reducing maternal mortality, the importance of having an experienced anaesthetist cannot be overemphasized.

Indications for elective caesarean sections Known cephalo-pelvic disproportion; placenta praevia; some malpresentations (p 126); after vaginal surgery (suburethral repair; vesico-vaginal fistula repair); or with some maternal infections, eg herpes, HIV (p 98). If a repeat section is planned arrange ultrasound placental localization to exclude placenta praevia as this is more common and more likely to be complicated by placenta accreta (hence risk of massive haemorrhage) in the presence of a uterine scar.

Emergency section may be needed because of antenatal complications, eg severe pre-eclampsia, abruptio placentae (baby still alive). In others, the need becomes apparent during labour: fetal distress; prolapsed cord (fetus alive); after failed induction; or failure to progress.

al of scar in labour: See p 134. Beware oxytocin. *Antibiotic prophylaxis* is commended. Infection (wound, endometritis, and UTI) is reduced by iv antibiotics (eg 2g cefradine at induction, 1g at 6h and 12h post-op) for both emergency and elective sections. Longer courses do not appear to be superior, nor do more expensive second generation cephalosporins.

After caesarean section In Rh-ve mothers, remove all excess blood from peritoneal cavity. Use a Kleihauer test (p 109) to determine dose of anti-D. Mobilize the mother early. If purported cephalo-pelvic disproportion led to section, consider pre-discharge radiographic pelvimetry.

Complications

- PPH
- Retained placental tissue
- Ureter trauma
- Vesico-uterine fistula
- Fetal injury (eg fracture)
- Colon injury

Prophylaxis against thromboembolism *Low-risk:* Women with no risk factors undergoing elective caesarean section in an uncomplicated pregnancy require only early mobilization and good hydration.

Moderate risk:

- Emergency section in labour
- Age >35 years
- Gross varicose veins
- Obesity (>80kg)
- Current infection
- Para 4+
- ≥4 days prior immobility
- Pre-eclampsia
- Major current illness

These women require heparin prophylaxis or mechanical methods.

High-risk: These women should all receive heparin until 5 days post-operatively until fully mobilized if longer. Leg stockings confers additional benefit.

Any woman with 3 risk factors above

Extended surgery, eg caesarean hysterectomy

On or past history of thromboembolism or thrombophilia

Paralysed lower limbs

Women with antiphospholipid antibody (cardiolipin antibody or lupus anticoagulant)

Women who have had thromboembolism in pregnancy should receive thromboprophylaxis for 6 weeks postpartum. See page 81.

►► Uterine rupture

Ruptured uterus is rare in the UK (1:1500 deliveries—but 1:100 in parts of Africa). Associated maternal mortality is 5%, and the fetal mortality 30%. ~70% of UK ruptures are due to dehiscence of caesarean section scars. Lower segment scars are far less likely to rupture (1.4%) than the classical scars (6.4%)—see p 132. Other risk factors: ●Obstructed labour in the multiparous, especially if oxytocin is used ●Previous cervical surgery ●High forceps delivery ●Internal version ●Breech extraction. Rupture is usually during the third trimester or in labour. **Trial of scar:** In general, a trial of labour is safer than automatic repeat caesars—particularly if the thickness of the lower uterine segment is >3.5mm on ultrasound at 36–38 weeks. Risk of subsequent rupture after caesarean is: 1.6/1000 if repeat caesarean without labour; 5.2/1000 where spontaneous labour occurs; 24.5/1000 if labour is induced using prostaglandins.¹ 20–40% undergo repeat emergency section.

Signs and symptoms Rupture is usually in labour. In a few (usually a caesarean scar dehiscence) rupture precedes labour. Pain is variable, some only having slight pain and tenderness over the uterus. In others pain is severe. Vaginal bleeding is variable and may be slight (bleeding is intraperitoneal). Unexplained maternal tachycardia, sudden maternal shock, cessation of contractions, disappearance of the presenting part from the pelvis, and fetal distress are other presentations. Postpartum indicators of rupture: continuous PPH with a well-contracted uterus; if bleeding continues postpartum after cervical repair; and whenever shock is present.

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Management If suspected in labour, perform laparotomy, deliver the baby by caesarean section, and explore the uterus. ►►●Give O₂ at 15L/min via a tight-fitting mask with reservoir ●Set up ivi ●Crossmatch 6u of blood and correct shock by fast transfusion. ●Arrange laparotomy. The type of operation performed should be decided by a senior obstetrician; if the rupture is small, repair may be carried out (possibly with tubal ligation); if the cervix or vagina are involved in the tear, hysterectomy may be necessary. Care is needed to identify the ureters and exclude them from sutures. Give post-operative antibiotic cover, eg ampicillin 500mg/6h iv and netilmicin 150mg/12h iv (unless there is renal impairment). 85% of spontaneous ruptures require hysterectomy, but >66% of ruptured scars are repairable.

►► Mendelson's syndrome

This is the name given to the cyanosis, bronchospasm, pulmonary oedema and tachycardia that develop due to inhalation of acid gastric contents during general anaesthesia. Clinically it may be difficult to distinguish from cardiac failure or amniotic fluid embolus. Pre-operative H₂ antagonist, sodium citrate, gastric emptying, cricoid pressure (p 770), the use of cuffed endotracheal tubes during anaesthesia, and pre-extubation emptying of the stomach aim to prevent it (p 132).

Management ►►Tilt the patient head down. Turn her to one side and aspirate the pharynx. Give 100% oxygen. Give aminophylline 5mg/kg iv, slow ivi and hydrocortisone 1000mg iv stat. The bronchial tree should be sucked out using a bronchoscope under general anaesthesia. Antibiotics, ampicillin and netilmicin (as above), should be given to prevent secondary pneumonia. Ventilation conducted on intensive care may be needed. Physiotherapy should be given during convalescence.

Abdominal pain in pregnancy¹

➤ With any pain in pregnancy think, could this be the onset of labour?

Abdominal pain may be from ligament stretching or from symphysis pubis strain. In early pregnancy remember miscarriage (p 22) and ectopics (p 24).

Abruption The triad of abdominal pain, uterine rigidity, and vaginal bleeding suggests this. It occurs in between 1 in 80 and 1 in 200 pregnancies. Fetal loss is high (up to 60%). A tender uterus is highly suggestive. Ultrasound may be diagnostic (but not necessarily so). A live viable fetus merits rapid delivery as demise can be sudden. Prepare for DIC, which complicates 33–50% of severe cases, and beware PPH, which is also common. See p 108.

Uterine rupture See opposite, p 134.

Uterine fibroids For torsion and red degeneration, see p 39.

Uterine torsion The uterus rotates axially 30–40° to the right in 80% of normal pregnancies. Rarely, it rotates >90° causing acute uterine torsion in mid or late pregnancy with abdominal pain, shock, a tense uterus, and urinary retention (catheterization may reveal a displaced urethra in twisted vagina). Fibroids, adnexal masses, or congenital asymmetrical uterine anomalies are present in 90%. Diagnosis is usually at laparotomy. Delivery is by caesarean section.

Ovarian tumours Torsion, rupture, see p 44. **Pyelonephritis** See p 160.

Appendicitis Incidence: ~1/1000 pregnancies. This is not commoner in pregnancy but mortality is higher (especially from 20 weeks). Perforation is commoner (15–20%). Fetal mortality is ~1.5% for simple appendicitis; ~30% when there is perforation. The appendix migrates upwards, outwards and posteriorly as pregnancy progresses, so pain is less well localized (often paraumbilical or subcostal—though right lower quadrant still commonest)² and tenderness, rebound, and guarding less obvious. Peritonitis can make the uterus tense and woody-hard. Operative delay is dangerous. Laparotomy over site of maximal tenderness with patient tilted 30° to the left should be performed by an experienced obstetric surgeon.

Cholecystitis Incidence 1–6 per 10,000 pregnancies. Pregnancy encourages gallstone formation due to biliary stasis and increased cholesterol in bile. Symptoms are similar to the non-pregnant with subcostal pain, nausea, and vomiting. Jaundice is uncommon (5%). Ultrasound confirms the presence of stones. The main differential diagnosis is appendicitis, and laparotomy or laparoscopy is mandatory if this cannot be excluded. Surgery should be reserved for complicated non-resolving biliary tract disease during pregnancy as in >90% the acute process resolves with conservative management. For patients requiring surgery, laparoscopic cholecystectomy can be a safe and effective method of treatment. medline:9448609

Rectus sheath haematoma Very rarely, bleeding into the rectus sheath and haematoma formation can occur with coughing (or spontaneously) in late pregnancy causing swelling and tenderness. Ultrasound is helpful.

ΔΔ: Abruption. **Management:** Laparotomy (or perhaps laparoscopy) is indicated if the diagnosis is in doubt or if there is shock. medline:11486328 & 9217951

Abdominal pain may complicate pre-eclampsia by liver congestion. Rarely, in severe pre-eclampsia the liver perforates. Pancreatitis in pregnancy is rare; but mortality high (37% maternal; 5.6% fetal). Diagnose by urinary diastase in first trimester when amylase may be low.¹

Stillbirth

Stillbirths are those babies born dead after 24 weeks completed gestation. Rate: 1:730 singleton pregnancies at 38 weeks. Death *in utero* may take place at any stage in pregnancy or in labour. Delivery is an emotional strain for both mother and attendant staff as the pain and process of labour may seem so futile and mothers may feel guilty—and that it is a punishment.

Some hours after a fetus has died *in utero* the skin begins to peel. On delivery such fetuses are described as *macerated*, as opposed to *fresh* stillbirths. If left, spontaneous labour usually occurs (80% within 2 weeks, 90% within 3 weeks), but it is common practice to induce labour once death is diagnosed, to prevent a long wait for labour for the mother, and minimize risk of coagulopathy. DIC (p 142) is rare unless a dead fetus of more than 20 weeks' gestation has been retained for >3 weeks. If tests indicate a clotting disorder is present, some authorities recommend heparin (1000u iv/h for up to 48h prior to onset of labour)—but get expert advice. Stop any heparin prior to labour, or if it supervenes. Ensure excellent supply of freshest possible blood and fresh frozen plasma are available at delivery.

Causes of stillbirth Pre-eclampsia, chronic hypertension, renal disease, diabetes, infection, hyperpyrexia ($T^{\circ} >39.4^{\circ}\text{C}$), malformation (11% of macerated stillbirths and 4% of fresh have chromosomal anomalies), haemolytic disease, postmaturity. Abruptio and knots in the cord may cause death in labour. In 20% no cause is found.

Diagnosis The mother usually reports absent fetal movements. No heart sounds (using Pinard's stethoscope, Doppler, or cardiotocography). There is no fetal movement (eg heart beat) on ultrasound. It may help the mother to see lack of heart beat.

Management The mother may want to go home after diagnosis to reflect, collect things and make arrangements. Check maternal T° , BP, urine for protein, and blood clotting screen if fetus not thought recently demised. Labour is induced using prostaglandin vaginally; or extra-amniotically (dose varied with uterine response). Oxytocin iv used concomitantly risks uterine or cervical trauma so use *after* prostaglandin extra-amniotic infusion ceases. Oxytocin infusion alone (p 110) may be used for induction if the cervix is ripe (Bishop's score ≥ 4 , gestation ≥ 35 weeks). Amniotomy is traditionally contraindicated as it risks infection. Some use mifepristone orally before using prostaglandins to induce abortion.

Ensure good pain relief in labour (if epidural, check clotting tests all normal). Do not leave the mother unattended. When the baby is born wrap it (as with any other baby) and offer to the mother to see and to hold—if the mother so desires. A photograph may be taken for her to take home a lock of the baby's hair and palm-print given (keep in notes for later if she does not want them then). Unseen babies are difficult to grieve for. Naming the baby and holding a funeral service may help with grief.

Labour ward procedure (to try to establish cause) Thorough examination of the stillbirth, clinical photographs. Take time to talk to parents about how helpful a postmortem may be to them, in understanding what happened, and planning further pregnancies. If consent is refused, MR (may miss significant pathology) and cytogenetics (\pm small volumes of tissue for metabolic studies) may be acceptable but are less informative. Do placental histology, high vaginal swab for bacteriology, maternal & fetal blood for TORCH^{ec} screen (p 98), and mother's blood for Kleihauer test, p 109 (detects fetomaternal transfusion—a cause of unexplained stillbirth) and lupus anticoagulant, and chromosome analysis of fetal blood and skin

Helping parents after stillbirth

Offer lactation suppression (bromocriptine 2.5mg po on day 1, then 2.5mg/12h po for 14 days), but beware if pre-eclampsia. Give parents a follow-up appointment to discuss causes found by the above tests. Consider a domiciliary visit if parents prefer. Refer for genetic counselling if appropriate. In England, a *Certificate of Stillbirth* is required (issued by an obstetrician), and the parents are required to take it to the Registrar of Births and Deaths within 42 days of birth. The father's name only appears in the register if the parents are married, or if both parents make the registration.

The Registrar then issues a Certificate of Burial or Cremation which the parents then give to the undertaker (if they have chosen a private funeral—in which case they bear the cost of the funeral), or to the hospital administrators if they have chosen a hospital funeral—for which the hospital bears the cost. Parents are issued with a Certificate of Registration to keep which has the name of the stillborn baby (if named), the name of the informant who made the registration, and the date of stillbirth.

UK hospitals are directed by the Department of Social Security to offer 'hospital' funerals for stillbirths (arranged through an undertaker). If the parents offer to pay for this, the hospital may accept. The hospital should notify the parents of the time of funeral so that they may attend, if they wish. With hospital funerals a coffin is provided and burial is often in a multiple-occupancy grave in a part of the graveyard set aside for babies. The hospital should inform parents of the site of the grave. Graves are unmarked, so should the parents not attend the funeral and wish to visit later it is recommended that they contact the graveyard attendants for the grave to be temporarily marked. Parents may buy a single occupancy grave, if they wish, on which they can later erect a headstone. Hospitals can arrange cremations, but the parents pay for this and parents should be told that there are unlikely to be any ashes after cremation.¹

In addition to arranging a follow-up appointment with the obstetrician to discuss implications for future pregnancy, and the cause (if known) of the stillbirth, it is helpful to give parents the address of a local branch of an organization for bereavement counselling, eg SANDS.² Grief may take a long time to resolve (p 442) and parents may find it difficult to contact ordinary medical staff without the 'excuse' provided by asking about the baby's ailments.

Each maternity unit should have a bereavement counsellor to support the mother and father, and help guide them through the formalities.

In the UK statutory maternity pay and the maternity allowance and social security maternity payments are payable after stillbirth.

Note: stillbirth affects ~1:730 singleton pregnancies by 38 weeks: it is 4 times as common as sudden infant death syndrome (p 286).

Post-partum haemorrhage (PPH)

Primary PPH is the loss of greater than 500mL (definitions vary) in the first 24h after delivery. This occurs after ~6% of deliveries; major PPH (>1 litre) is 1.3%. Causes: uterine atony (90%), genital tract trauma (7%), clotting disorders—p 142 (3%). Death rate: 2/yr in the UK; 125,000/yr worldwide.

Factors predisposing to poor uterine contractions • Past history of atony with PPH. • Retained placenta or cotyledon. • Ether or halothane anaesthesia. • A large placental site (twins, severe rhesus disease, large baby), low placenta, overdistended uterus (polyhydramnios, twins). • Extravasated blood in the myometrium (abruption). • Uterine malformation or fibroids. • Prolonged labour. • Poor second stage uterine contractions. • Trauma to uterus or cervix. • Older mothers.

Management • Give oxytocin 5u slowly iv. • Call emergency ambulance unit (p 92)—if not in hospital. Give high-flow O₂ as soon as available. • Set up iv (2 large-bore cannulae). • Call anaesthetist (a cvp line may help guide fluid replacement, but not if it causes delay). • If shocked give Haemaccel or fresh blood of the patient's ABO and Rh group (uncrossmatched in emergency) fast until systolic BP >100mmHg and urine flows at >30mL/h (catheterize the bladder). • Is the placenta delivered? If it is, is it complete? If not, explore the uterus. • If the placenta is complete, put the patient in the lithotomy position with adequate analgesia and good lighting. Check for and repair trauma. • If the placenta has not been delivered but has separated, attempt to deliver it by controlled cord traction after rubbing up uterine contraction. If this fails, ask an experienced obstetrician to remove it under general anaesthesia. Beware renal shut-down.

If bleeding continues despite all the above, give 10 units of oxytocin in 500mL dextrose saline, eg at a rate of 15 drops/min. Bimanual pressure on the uterus may decrease immediate loss. Inform consultant. Check that blood is clotting (5mL should clot in a 10mL round-bottomed glass tube in 6min; formal tests: platelets, prothrombin ratio, kaolin-cephalin clotting time, fibrin degradation products). Explore the uterus for possible rupture. If uterine atony is the cause, and the circulation is still compromised, give carboprost 250µg (15-methyl prostaglandin F_{2α}) eg as Hemabate® 1mL deep IM (max 8 doses, each >15min (usually 90min) apart). SE: nausea, vomiting, diarrhoea, T°↑; (less commonly—asthma, BP↑, pulmonary oedema). It controls bleeding in ~88%. Rarely, uterine packing, internal iliac artery or uterine vessel ligation, or hysterectomy (possibly safest) is needed to stop bleeding. Ask a haematologist's advice on clotting factor replacement (fresh frozen plasma contains all of them; the cryoprecipitate has more fibrinogen, but lacks antithrombin III).

Secondary PPH This is excessive blood loss from the genital tract after 24h from delivery. It usually occurs between 5 and 12 days and is due to retained placental tissue or clot. Secondary infection is common. Uterine involution may be incomplete. If bleeding is slight and there is no sign of infection it may be managed conservatively—but heavier loss, the suggestion of retained products on ultrasound, or a tender uterus with an open os, requires exploration. Crossmatch 2 units of blood pre-operatively. Give antibiotics (eg ampicillin 500mg/6h iv, metronidazole 1g/12h PR) if there are signs of infection. Carefully curette the uterus (it is easily perforated at this stage). Send curettings for histology (excludes choriocarcinoma).

Managing those refusing blood transfusion in pregnancy

- Know maternal attitude to transfusion at booking.
- Give oral iron and folate to mother to maximize haemoglobin stores.
- Book for delivery where there are good facilities to deal with haemorrhage promptly (including facilities for hysterectomy).
- Arrange ultrasound to know placental site.
- Inform consultant when admitted in labour. Ensure experienced staff conduct labour. Give oxytocin as soon as the baby is delivered. Do not leave the mother alone for first hour post-delivery.
- Consultants should perform caesarean section if required.
- Haemorrhage should be dealt with promptly, and clotting disorders excluded early. Involve a consultant obstetrician early (to decide if hysterectomy may be needed), and a consultant anaesthetist (for help with fluid replacement and for use of intensive care facilities). Liaise with a consultant haematologist. Avoid dextran (adversely affects haemostasis), but Haemacel® is useful.
- Ensure the woman does not want to change her mind and receive a transfusion.
- Should the woman die of exsanguination both bereaved relatives and distressed staff should be offered support.

Retained placenta

Physiological third stage takes 30min. With the use of oxytocic drugs and delivery and controlled cord traction, ie active management, the third stage is complete in 10min in 97% of labours, and a placenta not delivered in 30min will probably not be expelled spontaneously. The danger with retained placenta is haemorrhage. **Associations:** Previous retained placenta; previous uterine surgery; preterm delivery; maternal age >35 years; placental weight <600g; pethidine use in labour; induced labour; parity >5.

Management If the placenta does not separate readily, avoid excessive cord traction—the cord may snap or the uterus invert. Check that the placenta is not in the vagina. Palpate the abdomen. If the uterus is well contracted, the placenta is probably separated but trapped by the cervix. Wait for the cervix to relax and release it. If the uterus is bulky, the placenta may have failed to separate. Rub up a contraction, put the baby to the breast (stimulates oxytocin production) or give more Syntometrine®, and empty the bladder (a full bladder causes atony). If the placenta still will not deliver, prepare to remove it manually (delay may precipitate a PPH).

Manual removal Set up ivi and crossmatch blood (eg 2u). Call the anaesthetist. The procedure can be done under epidural if *in situ*: Obtain consent. With the mother in lithotomy position, using aseptic technique, place one hand on the abdomen to stabilize the uterus. Insert the other hand through the cervix into the uterus. Following the cord assists finding the placenta. Gently work round the placenta, separating it from the uterus using the ulnar border of the hand. When separated it should be possible to remove it by cord traction. Check that it is complete. Give oxytocic drugs and start antibiotics, eg doxycycline 200mg stat, 100mg/24h and metronidazole 500mg/8h iv.

Rarely, the placenta will not separate (placenta accreta) and hysterectomy may be necessary—by a senior obstetrician.

►► Uterine inversion

Inversion of the uterus is rare. It may be due to mismanagement of the third stage, eg with cord traction in an atonic uterus (between contractions) and a fundal insertion of the placenta. It may be completely revealed or partial when the uterus remains within the vagina. Even without haemorrhage the mother may become profoundly shocked.

Management The ease with which the uterus is replaced depends on the amount of time elapsed since inversion, as a tight ring forms at the neck of the inversion. With an inversion noted early before shock sets in, replacement by hand may be possible. If shock has ensued, set up a fast ivi and infuse colloid or blood. Summon expert help. Under halothane anaesthesia to relax the uterus, hold the uterus in the vagina with one hand. Run two litres of warm 0.9% saline fast into the vagina through cystoscopy tubing (or with a funnel and tube) with an assistant holding the labia encircled tightly around the operator's arm to prevent the fluid running away. Running the fluid through a silastic ventouse cap held in the vagina improves the 'vaginal seal'. The hydrostatic pressure of the water should reduce the uterus. Once the inversion has been corrected, give ergometrine to contract the uterus and prevent recurrence. Prophylactic antibiotics are advisable.

►► DIC and coagulation defects

DIC in pregnancy is always secondary to stimulation of coagulation by procoagulant substance release in the maternal circulation. Known triggers are: retention of a dead fetus (of greater than 20 weeks' gestation which has been dead for >3 weeks); pre-eclampsia; placental abruption; endotoxic shock; amniotic fluid embolism; placenta accreta; hydatidiform mole; prolonged shock from any cause; acute fatty liver of pregnancy (p 158). *Pathogenesis.* Thromboplastins are released into the circulation, fibrin and platelets are consumed as intravascular clotting occurs. *Tests:* Kaolin-cephalin clotting time↑ (↓ factors II, V, VII), fibrinogen↓, fibrin degradation products↑. In situations where DIC is a possibility send blood for crossmatch, platelets, partial thromboplastin time or accelerated whole blood clotting time, prothrombin time, fibrinogen estimation, and fibrin degradation products. Preliminary results should be available in 30min. *Management:* Presentation may be as heavy bleeding and shock, and the first measures must be the correction of shock. ►► Give O₂ at 15 litres/min via a tight fitting mask with reservoir. Set up at least 1, preferably 2, wide-gauge IVs, take bloods as above, and give blood fast (group-compatible blood—available in 5–10min or O Rh-ve blood if desperate). Stored blood is deficient in clotting factors. Give fresh frozen plasma to normalize the kaolin-cephalin clotting time and the prothrombin time. Platelets are indicated with prolonged bleeding and low platelet count. Calcium is sometimes needed to counteract citrate in stored blood (eg 10mL of 10% calcium gluconate iv, eg after 6u of blood. Seek expert help from a haematologist. The condition is usually self-limiting if the stimulus can be removed. In the case of intrauterine death and abruption (p 108) removal of the uterine contents is the way to correct the stimulus, and this should be done as promptly as possible. *Mortality:* <1% if placental abruption; 50–80% if infection/shock.

Autoimmune thrombocytopenic purpura (AITP) Incidence 1–2/10,000 pregnancies. IgG antibodies cause thrombocytopenia (associated with increased bone marrow megakaryocytes) in the mother, and being able to cross the placenta, they cause thrombocytopenia in ~10% of fetuses. Exclude systemic lupus erythematosus in the mother (thrombocytopenia may be an early presentation; do DNA binding, *OHCM* p 410). Consider maternal HIV. If maternal platelets fall below $20 \times 10^9/L$ or $50 \times 10^9/L$ near delivery, give steroids. Rarely, splenectomy is necessary during pregnancy (ideally in the second trimester). Immunoglobulin IgG 0.4g/kg iv for 5 days is sometimes used near expected date of delivery, inducing maternal and fetal remission for up to 3 weeks but it is extremely expensive. Aim for non-traumatic delivery for both mother and baby. Neonatal platelet count may fall further in the first days of life, then gradually rise to normal over 4–16 weeks. Treatment is not needed unless surgery is contemplated. Maternal mortality due to AITP is now negligible, but fetal mortality remains (due to intracranial bleeding). Take cord blood at delivery. If platelets $<20 \times 10^9/L$ give baby IgG 1g/kg iv at birth. If platelets low at birth observe baby for 2–5 days as further falls likely.

Thrombocytopenia in pregnancy 1 Spurious (try citrated bottle).

2 Pregnancy-associated thrombocytopenia (benign gestational thrombocytopenia)—mild and self-limiting (platelets stay above $100 \times 10^9/L$).

3 Autoimmune thrombocytopenia.

4 Pre-eclampsia (platelets may fall early, preceding clotting abnormality).

5 DIC (above) and haemolytic uraemic syndrome (p 280)/thrombotic thrombocytopenic purpura (2 ends of a microangiopathic spectrum, *OHCM* p 274).

6 Folate deficiency.

7 Congenital (May-Hegglin anomaly, hereditary macrothrombocytopenia).

8 Marrow disease; hypersplenism.

Amniotic fluid embolism

This condition, with a mortality up to 80% presents with sudden dyspnoea and hypotension, heralded, in 20%, by seizures. 40% also develop DIC, and of those who survive the initial collapse 70% go on to develop pulmonary oedema (acute respiratory distress syndrome, ARDS, *OHCM* p 176). An anaphylactic type of response occurs to (possibly abnormal) amniotic fluid in the maternal circulation. *Presentation* is often at the end of the first stage of labour or shortly after delivery but can complicate amniocentesis, or termination of pregnancy, and has even occurred up to 48h postpartum. Previously it was said to be related to uterine hyperstimulation (multiparous short labour and use of oxytocin). This may be anecdotal. Risk increases with increasing maternal age.

Management ►► The first priority is to prevent death from respiratory failure. Give mask oxygen and call an anaesthetist urgently. Endotracheal intubation and ventilation may be necessary. Set up IVI lest DIC should supervene. Treatment is essentially supportive—important steps are detailed below. Diagnosis may be difficult: exclude other causes of obstetric shock (p 106).

- Cardiopulmonary resuscitation if indicated.

- Give highest available O₂ concentration. If unconscious ventilate and use 100% inspired O₂.

- Monitor for fetal distress.

- If hypotensive, give fluids rapidly IVI to increase preload. If still hypotensive consider inotropes: dobutamine (a better inotrope than dopamine), eg in a dose range of 2.5–10 µg/kg/min IVI may help.

- Pulmonary artery catheterization (Swan–Ganz catheter if available) helps guide haemodynamic management.

- After initial hypotension is corrected, give only maintenance requirements of fluid to avoid pulmonary oedema from acute respiratory distress syndrome. Transfer to intensive care unit as soon as possible.

- Treat DIC with fresh whole blood or packed cells and fresh frozen plasma. Use of heparin is controversial, there is insufficient data to warrant routine heparinization.

- Should the woman die, specifically request that the lungs be examined for the presence of amniotic squames (to confirm the diagnosis).

Birth injuries

► Give all babies with signs of trauma vitamin K 1mg IM at birth (unless already given as part of routine measures).

To the baby—Moulding: This is a natural phenomenon rather than an injury. The skull bones are able to override each other (p 86) to reduce the diameter of the head. Moulding is assessed by the degree of overlap of the overriding at the sutures. If moulding is absent, the cranial bones are felt separately. With slight moulding, the bones just touch, then they override but can be reduced; finally they override so much that they cannot be reduced. Excessive moulding during labour indicates cephalo-pelvic disproportion, and can result in intracranial damage.

Cephalhaematoma: This is a *subperiosteal* swelling on the fetal head, and its boundaries are therefore limited by the individual bone margins (commonest over parietal bones). It is fluctuant. Spontaneous absorption occurs but may take weeks and may cause or contribute to jaundice.

Caput succedaneum: This is an oedematous swelling of the scalp, superficial to the cranial periosteum (which does not, therefore, limit its extent) and is the result of venous congestion and exuded serum caused by pressure against the cervix and lower segment during labour. The presenting part of the head therefore has the swelling over it. It gradually disappears in the first days after birth. When ventouse extraction is used in labour a particularly large caput (called a chignon) is formed under the ventouse cup.

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Erb's palsy: This may result from a difficult assisted delivery, eg shoulder dystocia. The baby's arm is flaccid and the hand is in the 'porter's tip' posture (p 724). Exclude a fractured clavicle and arrange physiotherapy. If it has not resolved by 6 months, the outlook is poor.

Subaponeurotic haematoma: Blood lies between the aponeurosis and the periosteum. As haematoma is not confined to the boundaries of one bone, collections of blood may be large enough to result in anaemia or jaundice. They are associated with vacuum extractions.

Skull fractures: These are associated with difficult forceps extraction. They are commonest over parietal or frontal bones. If depressed fractures are associated with neurological signs, ask a neurosurgeon if the bone should be elevated.

Intracranial injuries: Intracranial haemorrhage is especially associated with difficult or fast labour, instrumental labour, and breech delivery. Premature babies are especially vulnerable. Normally a degree of motility of intracranial contents is buffered by cerebrospinal fluid. Excessive moulding and sudden changes in pressure reduce this effect and are associated with trauma. In all cases of intracranial haemorrhage check babies' platelets. If low, check mother's blood for platelet alloantibodies (PLA1 system). Subsequent babies are at equal risk. iv maternal immunoglobulin treatment is being evaluated.

Anoxia may cause intraventricular haemorrhage (p 232). Asphyxia causes intracerebral haemorrhage (often petechial) and may result in cerebral palsy. Extradural, subdural, and subarachnoid haemorrhages can all occur. Babies affected may have convulsions, apnoea, cyanosis, abnormal pallor, low heart rate, alterations in muscle tone, restlessness, somnolence, or abnormal movements. Treatment is supportive and expectant. See p 232 & p 234.

Birth injuries to the mother: anal sphincter injury

This injury is common, and is not completely preventable by episiotomy (p 147). Faecal incontinence is a source of misery, and requires expert attention. *Do not be tempted to sew up rectal tears unless you are sure they are superficial* (and then sewing may not be needed). When in doubt, get help.

Risk of mechanical injury is greatest after the first vaginal delivery. Traumatic stretching of the pudendal nerves occurs in >30% of primips, but is mostly asymptomatic, or mildly/transiently so. These patients are at ↑ risk in subsequent deliveries (cumulative pudendal nerve injury is well-recognized).

If rectal incontinence occurs, and especially if there is a recto-vaginal fistula, get expert surgical help.

M Fynes 1999 *Lancet* 354 983 [10501360]

Episiotomy and tears

Perineal tears These are classified by the degree of damage caused. Tears are most likely to occur with big babies, precipitant labours, babies with poorly flexed heads, shoulder dystocia, when forceps are used, or if there is a narrow suprapubic arch. Perineal massage in pregnancy helps prevent perineal trauma but may be uncomfortable at first.¹

Labial tears Common, these heal quickly and suturing is rarely helpful.

First degree tears These tears are superficial and do not damage muscle. They may not need suturing unless blood loss is marked.

Second degree tears These lacerations involve perineal muscle and severe may involve the anal sphincter. They are repaired in a similar fashion to repair of episiotomy (see below).

Third degree tears Damage extends through the anal mucosa and can extend into the rectum (see p 145). Repair by an experienced surgeon under epidural or general anaesthesia. Rectal mucosa is repaired first using catgut from above the tear's apex to the mucocutaneous junction. Muscle is interposed. Vaginal mucosa is then sutured. Severed ends of the anal sphincter are apposed using figure-of-eight stitches. Finally skin is repaired. Avoid constipation post-operatively by using a high-fibre diet and faecal softeners.

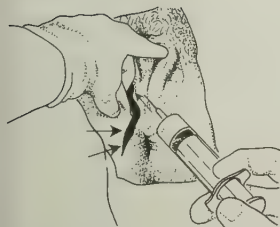
Episiotomy This is performed to enlarge the outlet, eg to hasten birth of a distressed baby, for instrumental or breech delivery, to protect a premature head, and to try to prevent 3° tears (but anal tears are not reduced by more episiotomies in normal deliveries). In some hospitals, the episiotomy rate is >90%, and there is no evidence to justify its use on this scale.¹

The tissues which are incised are vaginal epithelium, perineal skin, bulbospongiosus muscle, superficial, and deep transverse perineal muscles. With large episiotomies, the external anal sphincter or levator ani may be partially cut, and ischiorectal fat exposed.

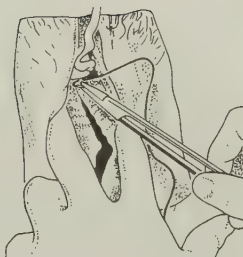
Technique: Hold the perineal skin away from the presenting part of the fetus (2 fingers in vagina). Infiltrate area to be cut with local anaesthetic, eg 1% lidocaine (=lignocaine). Still keeping the fingers in the introitus, cut medially laterally towards the ischial tuberosity, starting medially (6 o'clock), so avoiding the Bartholin's glands. (Midline episiotomy is ineffective at protecting perineum and sphincters and may impair anal continence.³)

Repair: (See diagrams.) NB: use resorbable sutures, but not glycerol-impregnated cat-gut (risks long-term dyspareunia). In lithotomy, and using good illumination, repair the vaginal mucosa first. Start above the apex using interlocking stitches 1cm apart, 1cm from wound edges. Tie off at mucocutaneous junction of fourchette. Then repair muscles with interrupted stitches to obliterate any dead spaces. Finally close the skin (subcutaneous stitch is more comfortable than interrupted stitches).

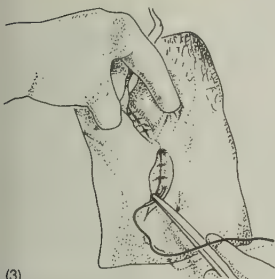
Problems with episiotomy: Bleeding (so may increase chance of spread of HIV from mother to baby); infection and breakdown; haematoma formation. For comfort some suggest ice packs, salt baths, hair dryer to dry perineum. 60% of women suffer perineal damage (episiotomy or tear) with spontaneous vaginal delivery; rectal diclofenac can provide effective analgesia. Superficial dyspareunia: see p 74. If labia minora are involved in the skin bridge, the introitus is left too small. If the deep layers are inadequately sutured, the introitus becomes rather rounded exposing the bladder to coital thrusts.



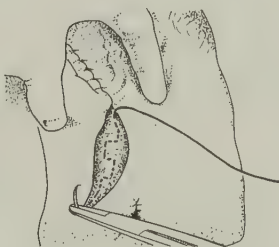
(1)
Swab the vulva towards the perineum. Infiltrate with 1% lignocaine → (arrows).



(2)
Place tampon with attached tape in upper vagina. Insert 1st suture above apex of vaginal cut (not too deep as underlying rectal mucosa nearby).

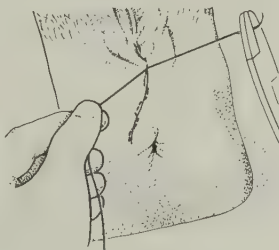


(3)
Bring together vaginal edges with continuous stitches placed 1 cm apart. Knot at introitus under the skin. Appose divided levator ani muscles with 2 or 3 interrupted sutures.



(4)
Close perineal skin (subcuticular continuous stitch is shown here).

(5)
When stitching is finished, remove tampon and examine vagina (to check for retained swabs). Do a PR to check that apical sutures have not penetrated rectum.



The puerperium

The puerperium is the 6 weeks after delivery. The uterus involutes, from 1kg weight at delivery to 100g. Felt at the umbilicus after delivery, it is a pelvic organ at 10 days. Afterpains are felt (especially while suckling) as it contracts. The cervix becomes firm over 3 days. The internal os closes by 3 days, the external os by 3 weeks. Lochia (endometrial slough, red cells, and white cells) is passed *per vaginam*. It is red (*lochia rubra*) for the 1st 3 days, then becomes yellow (*lochia serosa*) then white over the next 10 days (*lochia alba*), until 6 weeks. The breasts produce milky discharge and colostrum during the last trimester. Milk replaces colostrum 3 days after birth. Breasts are swollen, red, and tender with physiological engorgement at 3 to 4 days.

The first days If Rh-ve, give anti-D, within 72h, p 109. Check T°, BP, breasts, legs, lochia, fundal height. Teach pelvic floor exercises. Persistent red lochia, failure of uterine involution, or PPH (p 138) suggest retained products. Sustained hypertension may need drugs (OHCM p 126). ► Check rubella immunity. Vaccinate if non-immune but delay until postnatal exam if anti-D given. Check Hb on postnatal day 1 or ≥day 7: postpartum physiological haemodilution occurs from days 2–6.

Puerperal pyrexia is a temperature >38°C in the first 14 days after delivery or miscarriage. Examine fully (chest, breasts, legs, lochia, and bimanual vaginal examination). Culture msu, high vaginal swabs, blood, and sputum. 90% of infections will be urinary or of the genital tract. Superficial perineal infections occur around the second day. Endometritis gives lower abdominal pain, offensive lochia, and a tender uterus (on bimanual vaginal exam). Endometritis requires iv antibiotics (see below) and uterine curettage. For breast infection give flucloxacillin 250mg/6h po early, to prevent abscesses. Suckling or breast expression should continue to prevent milk stagnation. Even if the cause of pyrexia is not apparent, it is wise to treat with amoxicillin 500mg/8h po or iv and metronidazole 500mg/8h po. NB puerperal infection can be prevented by cleansing the birth canal at every vaginal examination with 0.25% chlorhexidine.

Superficial thrombophlebitis This affects 1% of women, presenting with a painful tender (usually varicose) vein. Give anti-inflammatories, eg ibuprofen 400mg/8h po pc. Bandage and elevate the leg. Recovery is usual within 4 days. Deep vein thrombosis: See p 151.

Puerperal psychosis (1 : 500 births): ► See p 408. This is distinguished from the mild depression that often follows birth by a high suicidal drive, severe depression (p 336), mania, and more rarely schizophrenic symptoms (p 358) with delusions that the child is malformed. If an acute organic reaction (p 350) is present, suspect puerperal infection. It may be worthwhile explaining to mothers that the puerperium is not always a time of joy, and they may feel low—and should let the midwife, health visitor or GP know about negative feelings, persistent crying, sleeplessness, and feelings of inadequacy—as early recognition of the problem is the best way to avoid what can be an intensely destructive experience. *Treatment:* See p 408 for a fuller discussion.

The 6-week postnatal examination gives a chance to: • See how mother and baby relate. • Do BP & weight. • Do FBC if anaemic postnatally. • Arrange a cervical smear if not done antenatally. • Check contraceptive plans (p 66) are enacted (start Pills 3 weeks postnatally). • Ask about depression, backache, incontinence. Ask 'have you resumed intercourse?' Sexual problems are common, and prolonged: ~50% report that intercourse is less satisfactory than pre-pregnancy, with major loss of libido, and dyspareunia the chief complaints. * Vaginal examination to check healing is not usually needed.

Venous thromboembolism (VTE) in pregnancy

►Investigate any unexplained calf or chest symptoms **today**. Thromboembolism is the chief UK cause of maternal death. VTE risk rises 6-fold in pregnancy (~0.3–1.6% of pregnancies: 20–50% occur antenatally—in any trimester). For those at special risk see p 81. 50% of those with 1st VTE have thrombophilia (p 151), so check for this. Where clinical suspicion is strong, but tests negative, start treatment, stop only after repeat negative test 1 week later.¹ Before starting heparin take blood for FBC, U&E, LFTs, coagulation and thrombophilia screens.

Pulmonary embolism Small emboli may cause unexplained pyrexia, syncope, cough, chest pain, and breathlessness. Pleurisy should be considered due to embolism unless there is high fever or much purulent sputum. Large emboli present as collapse with chest pain, breathlessness, and cyanosis. There will be a raised JVP, third heart sound, and parasternal heave.

Tests: CXR and ECG may be normal (apart from showing a tachycardia, the most consistent finding). ECG changes of deep S-wave in lead I and Q-wave and inverted T-wave in lead III can be caused by pregnancy alone. Blood gases may be helpful ($P_aO_2 \downarrow$; $P_aCO_2 \downarrow$). CT pulmonary angiograms are better than ventilation/perfusion scans. Test for venous thrombi (eg plasma D-dimer).

Treatment: Massive emboli may require prolonged cardiac massage and consideration of pulmonary embolectomy. Give heparin 5000u iv, (10,000u in severe pulmonary embolism), then ~1600u/h iv in 0.9% saline by syringe pump. Monitor APTT (OHCM p 650). After 3–7 days on iv heparin maintain on long-term heparin (eg 10,000u/12h sc with careful monitoring. Keep level <0.2u/mL. Problems: maternal osteopaenia (reversible on stopping); thrombocytopenia; allergic skin rashes; alopecia. For less massive emboli, low molecular weight heparin eg enoxaparin, (see below), is an alternative. Warfarin is teratogenic (Conradi–Hünemann syndrome, p 744) and is used antenatally up to 36 weeks only in those with artificial heart valves. sc heparin is continued through labour with no increased risk of PPH, but epidural anaesthesia is contraindicated. Heparin postpartum is reduced to 7500u/12h sc. Treat for at least 6 weeks post-partum and 3 months from embolus. Some choose warfarin use from 2 days postpartum.

Deep vein thrombosis (DVT) DVTs typically present with calf pain. For other signs, see OHCM p 444. Often symptoms are not clear-cut. Tests: Doppler ultrasound. **Treatment:** Elevate leg and use compression stockings. Give heparin throughout pregnancy eg enoxaparin 1mg/kg/12h sc (based on early pregnancy weight); measure peak anti-Xa activity 3h post-injection aim for level between 0.6–1u/mL (if above this reduce dose eg from 80mg/12h to 60mg/12h and repeat anti-Xa). Check platelets at 7 days and then monthly (if heparin-induced thrombocytopenia occurs use danaparoid sodium). After delivery warfarin may be used (breast feeding is ok). Monitor INR meticulously. Treat for at least 6 weeks post-partum and 3 months from thrombosis. Compression stockings should be worn for 2 years.¹

Prophylaxis See pages 81, and 133. Heparin is used for high-risk patients: ie in recurrent thromboembolism or thromboembolism associated with thrombophilia (p 151). Enoxaparin 40mg daily seems efficacious and safe. Lower risk women (previous single thromboembolic episode) receive heparin 7500u sc during labour, induction, or caesarean section (after taking plenty of blood in case crossmatch is needed). Then use heparin for 2 weeks post-delivery or heparin then warfarin (as above).

Thrombophilia in pregnancy^{1,2}

Thrombophilia is a tendency to increased clotting and many underlying causes are now known to contribute. Collectively, the conditions below affect at least 15% of Western populations; but are found in 50% of those with episodes of venous thromboembolism. Factors include:

- Factor V Leiden mutation: This ↓ factor V breakdown by protein C (activated protein C (APC) resistance). This affects 4–10% of the population and increases thrombotic risk 3–8 times. Homozygous individuals have 80 times the risk of venous thromboembolism.
- Protein C deficiency (affects 0.3% of population; increasing thrombotic risk 10–15 times).
- Protein S deficiency (affects 2% of population; ↑ thrombotic risk 10 times).
- Antithrombin III deficiency (affects 0.02% of population; thrombotic risk is increased 25–50 times).
- Lupus anticoagulant ± cardiolipin antibody. Women with lupus anticoagulant risk arterial and venous thrombosis; in atypical veins, eg portal or arm.
- G20210A mutation of the prothrombin gene (2% of the population).
- Homozygosity for the thermolabile variant of methylenetetrahydrofolate reductase (c677T MTHFR), which leads to homocysteinaemia (10% of the population).
- Dysfibrinogenaemia is rare, and the thrombotic risk variable.

Pregnancy is an acquired risk factor for venous thromboembolism. The difficulty is to know who has thrombophilia; and what risk this poses to a pregnant woman. Retrospective work from pregnancies complicated by thrombophilia (n=72,000), suggested risk of thromboembolism was 1:437 for women with factor V Leiden; 1:113 among those with protein C deficiency; 1:2.8 in those with type 1 (quantitative) antithrombin deficiency, and 1:42 amongst those with type 11 (qualitative) antithrombin deficiency. No association was found between venous thromboembolism and homozygosity for c677T MTHFR mutation, perhaps because plasma homocysteine reduces in pregnancy and folic acid supplements ameliorate hyperhomocysteinaemia.

Another study looking at relative risk of thromboembolism during pregnancy and postpartum suggests relative risk of thromboembolism is 9.3 for those with factor V Leiden; 15.2 for those with G20210A prothrombin gene mutation; relative risk for those affected by both was 107. By assuming an underlying rate of thromboembolism of 0.67 per 1000 pregnancies expected rates of thromboembolism of 1:500 was estimated for those with factor Leiden; 1:200 for those with G20210A prothrombin gene mutation and 4.6:100 for those with both. Homozygosity for the c677T MTHFR mutation was not associated with increased risk.

It is not felt that all women should be screened for thrombophilia; although screening is recommended for those with a family history of thromboembolism and those with past history of thromboembolism. Screening is also recommended in those with second-trimester pregnancy loss; severe or recurrent eclampsia and intrauterine growth restriction although it is not yet known how knowledge of thrombophilia will affect management. Those with thrombophilia but without previous history of thromboembolism require individualized assessment of the defect and additional risk factors by clinicians with expertise in this area. Drug (eg low molecular weight heparin), treatment is recommended for those with thrombophilia and past history of thromboembolism.

Cardiac disease in pregnancy

During pregnancy cardiac output increases to a maximum of 30–40% above non-pregnant levels. This is effected by ↑ heart rate and stroke volume.

Heart disease affects 0.5–2% of pregnant women. Examine the heart carefully early in all pregnancies. Ask the opinion of a cardiologist if there is doubt: ● Past history (eg congenital heart disease, rheumatic fever). ● Previous Kawasaki disease (now a more common cause of acquired heart disease than rheumatic fever). ● Murmurs (other than 5–7 in the list below).

60% of maternal cardiac deaths occur after delivery. Cardiac failure can occur at any stage in pregnancy but risk increases as pregnancy advances and is greatest in the early puerperium. Eisenmenger's syndrome risks maternal mortality (MM) of 30%; pulmonary hypertension (MM=30–50%); coarctation of the aorta (surges of BP↑ in the proximal segment), severe aortic or mitral stenosis and inoperable cyanotic heart disease are associated with ↑MM, so advise against pregnancy—or arrange meticulous specialist care. Seek pre-pregnancy advice for those with Marfan's. Termination may be medically advised.

Prosthetic valves with anticoagulation: Get expert help. Warfarin use risks fetal loss (p 744); heparin use risks maternal haemorrhage. medline:10688509

Antenatal management Regular visits to cardiologist/obstetric combined clinic. Prevent anaemia, obesity, and smoking. Ensure sufficient rest. Treat hypertension. Treat infections early. Give antibiotic cover for dental treatment (OHCM p 138). Examine carefully to exclude pulmonary oedema and arrhythmias at all visits. Heart failure requires admission.

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Labour If cardiac reserves are good before labour, risks during labour are low. Have oxygen and drugs to treat cardiac failure to hand. Avoid lithotomy position (dangerous ↑venous return after labour—the best position is semi-sitting). Aim for vaginal delivery at term with a short second stage (lift out forceps or ventouse). Give antibiotic cover, eg ampicillin 500mg iv and gentamicin 80mg iv repeated twice at 8-hourly intervals, if valve or septal defect. Pain relief should be good. Epidurals are safe if hypotension is avoided. Beware large volumes of iv fluids. Avoid ergometrine (use syntocinon, if necessary). Caesarean section should not be attempted (except during eclampsia) if the patient is in failure. Cardiac failure is most likely within the first 24h after delivery, so ensure very careful observations at this time.

Cardiac failure If symptoms or signs detected, admit the mother for bed rest, and treatment with diuretics ± digoxin. If acute failure develops, give 100% oxygen, nurse semi-recumbent, and give furosemide (=frusemide) 40mg iv slowly (<4mg/min), morphine 10mg iv, and aminophylline 5mg/kg by slow ivi (over >15min). If there is no improvement, consider ventilation. (There is no specific information on the use of nitrates in labour.)

Arrhythmias *Atrial fibrillation:* Is there mitral stenosis too?—admit to hospital. Treatment is as for the non-pregnant and may include digitalization or cardioversion. *Supraventricular tachycardia:* This may precipitate cardiac failure. If Valsalva manoeuvre and carotid massage fail, anaesthetize and cardiovert.

These signs may be normal in pregnancy 1 Oedema; ↑ pulse volume.

2 Vigorously pulsating neck veins (but JVP should not be ↑).

3 The apex beat is forceful (but <2cm lateral to midclavicular line).

4 The first heart sound is loud and a third heart sound can be heard in 84%.

5 An ejection systolic murmur is heard in 96% of women.

6 Systolic or continuous murmurs over either 2nd intercostal spaces 2cm from sternal edge, modified by pressure may be from mammary vessels.

7 Venous hums may be heard in the neck (modified by posture).

8 CXR may show slight cardiomegaly, ↑ pulmonary vascular markings, distension of pulmonary veins due to ↑ cardiac output.

Psychopharmacology in pregnancy

For puerperal depression, see p 408.

We always try to avoid drugs in pregnancy, but sometimes psychotropics are essential, eg if the mother is neglecting herself or her pregnancy. When psychotropic drugs are used, they must be combined with well-planned psychosocial support from a trusted confidant of the patient.

Depression in pregnancy—*Unipolar*: For diagnosis, see p 336. Try to wait until the second trimester before prescribing, and avoid SSRIs (p 340) as there is little experience with their use. Most experience is with tricyclics (p 340) such as amitriptyline—which are therefore the first choice when drugs are essential, despite warnings in naturally cautious *Data sheets* that read ‘...The safety of amitriptyline has not been established in pregnancy. It is not recommended during pregnancy, especially during the first and third trimesters unless there are compelling reasons, and, in these patients, the benefits should be weighed against the possible hazards to mother, fetus, and child... Animal experiments have shown harmful effects at exceptionally high doses...’ For these reasons, it is wise to discuss decisions with another doctor, or your local drug information service. Problems are unlikely, but the exact risk of teratogenesis cannot be stated. Withdrawal effects have been noted in neonates, eg agitation and respiratory depression with amitriptyline, and colic, spasms, and hyper- or hypotension with imipramine. Get a second opinion if ECT may be indicated. In general, breast feeding is contraindicated, as metabolites pass to the baby.

Bipolar: Lithium (Li^+ , p 354) is linked with teratogenicity (eg displacement of the tricuspid valve into the right ventricle). Offer specialist fetal echocardiography at 16 weeks in those women electing to remain on Li^+ . Used outside the first trimester, lithium can still cause problems with the fetal renal, and thyroid function. Monitor drug levels frequently (exactly 12h post-dose), and keep the dose as low as possible, and, in general, aim for a level of $<0.4\text{mmol/L}$. Do not change brands, as bioavailability varies. Do extra monitoring during intercurrent illness, D&V, and when poor compliance or toxicity is suspected (tremor, drowsiness, visual disturbance).

If lithium is stopped for a pregnancy or labour, restart it within a few days of birth. It is excreted in breast milk, so breast feeding is contraindicated.

Phenothiazines in pregnant schizophrenics There is conflicting data on safety—but there is agreement that most pregnancies will be unaffected. Pre-birth exposure may result in a syndrome of hyperreflexia, hypertonia, and tremor, which may persist for the first months of life. NB: Rates of fetal abnormality are increased in schizophrenia, even in those taking no drugs. The *Data sheet* for chlorpromazine reads ‘... There is inadequate evidence of safety in human pregnancy but it has been widely used for many years without apparent ill consequence. There is evidence of harmful effects in animals... It should be avoided in pregnancy unless the physician considers it essential. It may occasionally prolong labour, and at such a time should be withheld until the cervix is dilated 3–4cm. Possible adverse effects on the neonate include lethargy or paradoxical excitability, tremor, and low Apgar score. Being excreted in milk, breast feeding during treatment should be suspended...’

Anxiety in pregnancy Temazepam has been used successfully in those where insomnia has been a real problem. Avoid diazepam around the time of delivery, as withdrawal may occur in the baby (floppy baby syndrome). Avoid β -blockers, as these retard fetal growth. Relaxation techniques (p 344) and supportive psychotherapy (p 376) are far more appropriate.

Anaemia in pregnancy

► Even a small PPH may become life-threatening if the mother is anaemic. Anaemia predisposes to infection, and makes heart failure worse. Worn-out anaemic mothers may not cope with their offspring. Anaemia is the main cause of perinatal problems associated with malaria; above all, anaemia is the leading mechanism by which poverty exacts its morbid toll in pregnancy.

WHO definition of anaemia of pregnancy Hb <11g/dL. By this standard 50% of women not on haematinics become anaemic. The fall in Hb is steepest around 20 weeks' gestation, and is physiological (p 80); indeed failure of Hb to fall below 10.5g/dL (but not further than ~9.5g/dL) indicates ↑ risk of low birth weight or premature delivery.

Who is prone to anaemia? Those who start pregnancy anaemic, eg from menorrhagia, hookworm, malaria, with haemoglobinopathies; those with frequent pregnancies, twin pregnancy, or a poor diet.

Antenatal screening includes Hb estimation at booking, at 28 and 36 weeks. In black patients do sickle-cell tests, in others of foreign descent consider Hb electrophoresis for other haemoglobinopathies. From malarious areas consider malaria, and thick films. See p 159.

Treatment Pregnancy ↑ iron needs by 700–1400mg (per pregnancy), provided for by a pregnancy-induced 9-fold increase in iron absorption. Iron and folate supplements (and prevention against hookworm and malaria) are recommended in developing countries.

medline:1075925

154 Offer iron to those likely to be iron deficient (see above) or who would refuse transfusion if haemorrhaging (p 139). **Parenteral iron** may be given (to those with iron deficiency anaemia not tolerating oral iron) as iron sorbitol citric acid 1.5mg/kg IM daily (max 100mg/injection) as per *Data Sheet* >24h after stopping oral iron to prevent toxic reaction of headache, nausea, and vomiting. Haemoglobin rises over 6 weeks, so late severe anaemia (Hb <9g/dL) may need blood transfusion. One unit of blood increases the Hb by ~0.7g/dL.

Thalassaemia These globin chain production disorders are found in Mediterranean, Indian, and South-east Asian populations. Although anaemic never give parenteral iron as iron levels are high. Seek expert advice as to use of oral iron and folate. β -Thalassaemia does not affect the fetus but in homozygotes regular transfusions sustain life only until young adulthood. There are α chains in fetal HbF, so in α -thalassaemias the fetus may be anaemic or, if severe, stillborn. Mothers carrying lethally affected hydropic fetuses risk severe pre-eclampsia, and delivery complications due to a large fetus and bulky placenta. Prenatal diagnosis is possible by chorionic villus sampling (p 166) for thalassaemias anticipated by parental blood studies.

Sickling disorders can affect people of African origin, Saudi Arabians, Indians, and Mediterranean populations. **Sickle-cell trait** is not usually a problem. **Sickle-cell disease** predisposes to abortion, preterm labour, stillbirth, crisis. There is a chronic haemolysis (eg Hb 6–9g/dL). Regular 3–4L blood transfusions every 6 weeks (so a problem is development of atypical antibodies) prevents crises. Infection may induce crises, and dehydration exacerbates them; treat with exchange transfusions (OHCM p 642). **Sickle-cell haemoglobin C disease** is a milder variant of sickle-cell disease. Hb levels usually near normal so women may be unaware they are affected. They are still susceptible to sickling crises in pregnancy and the puerperium, so antenatal diagnosis is essential. Prenatal sickle-cell diagnosis is possible by chorionic villus sampling.

► Aim for diagnosis at birth (cord blood) at the latest so that penicillin pneumococcal prophylaxis may be started (OHCM p 642).

HIV in pregnancy and labour

In sub-Saharan Africa HIV is common (prevalence ~24% in apparently well antenatal patients—and there are millions of AIDS orphans). In Zimbabwe 120,000 HIV+ve mothers give birth each year. Many babies are not infected *in utero* but become so during parturition. Most mothers at risk of passing on HIV to the next generation (vertical transmission) do not want to know their HIV status, or cannot afford to find out. (If their husband is uninfected, what is the chance of him remaining loyal?) In our section on perinatal HIV (p 98) we comment that zidovudine (AZT) and caesarean section can prevent vertical transmission—but this is not much help if the nearest hospital is 3h away by wheelbarrow, and has only basic drugs. What is needed is much more cost-effective advice.

- Only give blood transfusions if absolutely necessary.
- Avoid any procedure likely to lead to maternal cells contacting fetal blood—eg external cephalic version, and amniocentesis.
- In instrumental deliveries, try to avoid abrasions of the fetal skin. Vacuum extractors may be preferable to forceps.
- When you clamp the cord, ensure there is no maternal blood on it.
- Artificial rupture of membranes and episiotomies should be left to the last possible moment, or avoided altogether.
- Avoid fetal scalp electrodes, and doing fetal scalp blood samples.
- If the membranes have ruptured, and labour is delayed or slow, consider the use of oxytocin to decrease exposure of the fetus to HIV.
- During caesarean sections, open the last layer by blunt dissection, to avoid minor cuts to the baby from the scalpel.
- Rinse the baby after birth; wipe the face away from mouth, eyes, and nostrils.
- Unless there is apnoea, avoid suction catheters to aspirate mucus from the nostrils. The baby's face is likely to be covered with the mother's blood at this stage, and you do not want to force HIV into nostrils.
- Health programmes are likely to end up encouraging breast feeding if there is no satisfactory alternative. Humanized milk is expensive, and may indicate to the mother's neighbours that she is HIV+ve. Using humanized milk might also compound problems by removing the one free method of contraception: lactational amenorrhoea. But studies from Nairobi showed 70% formula fed babies alive and disease-free at 2yrs vs 58% if breast fed. Most transmission occurred in the first 6 months. These babies had access to the city water supply.¹ Breast milk transmission was 16%. Also HIV+ve mothers in developing countries who breast feed may die sooner.²
- Offer advice on avoiding future pregnancies. This is not an easy area. Encouraging the use of condoms is fine, but many will want the added protection of the Pill. IUDs promote bleeding, and may increase spread to men. This may also be a problem with the progestogen-only pill, but note that the latter may cause less ectropion than the combined pill, and this might be advantageous. Sterilization is the hardest choice, especially when the mother now has no living children because of HIV.

Traditionally, drug therapy for HIV in pregnant patients has consisted of monotherapy with zidovudine—in contrast to non-pregnant women who are best treated with combination therapy (other agents include ritonavir and ddi, see *OHCM* p 586). Zidovudine monotherapy *may* be appropriate as the sole treatment in HIV +ve women with undepleted CD4 counts and low viral loads. In other circumstances, such treatment is likely to be suboptimal. Get expert advice. If a patient on triple therapy falls pregnant, most experts would continue treatment.

Diabetes mellitus (DM) in pregnancy

► Meticulous control around conception ↓ malformation rates. Tell all diabetics about preconception services; they must know *before* pregnancy. DM may be pre-existing or appear in pregnancy; glycosuria unrelated to DM is common (glomerular filtration ↑ and tubular glucose reabsorption ↓). Maternal glucose levels in pregnancy are normally constant (4–4.5 mmol/L) except after meals. Some screen at 1st antenatal visit with a blood sample 1h after a 50g glucose loaded fruit drink and do a full 75g oral glucose tolerance test (OGTT, *OHCM* p 283) and HbA_{1c} if the plasma glucose is ≥ 7 mmol/L.

Complications *Maternal:* Polyhydramnios (25%—?due to fetal polyuria), preterm labour (17%—associated with polyhydramnios). Stillbirth near term was common. *Fetal:* Malformation rates ↑ 3–4-fold. Sacral agenesis, almost exclusive to diabetic offspring, is rare (CNS & CVS malformations are much commoner). Babies may be macrosomic (too large) or sometimes growth restricted. *Neonatal risks:* Hypoglycaemia, Ca²⁺ ↓, Mg²⁺ ↓, and RDS (p 240). They may be polycythaemic (so more neonatal jaundice).

Antenatal care Liaise with diabetologist. Confirm gestation with early ultrasound. Detailed abnormality scan at 18–20 weeks. Fetal echo at 24 weeks. Educate about benefits of normoglycaemia and home glucose monitoring; regular postprandial monitoring *does* prevent harm to the baby. Insulin needs increase by 50–100%. One approach to normoglycaemia uses Actrapid® 20 minutes before meals with a Novopen® as described on p 262) with a single daily dose of Human Ultratard®. Admit unless there is good control at home. Oral hypoglycaemics are currently avoided although glyburide does not cross the placenta and may be safe. medline:11036125

Monitor fetal growth and wellbeing by ultrasound and cardiotocography.

Delivery Timing takes into account control of diabetes, any pre-eclampsia, maturity and size of the baby, and with attention to fetal wellbeing. Delivery before 38 weeks may result in neonatal respiratory distress. Deliver the baby where there are good neonatal facilities. Traditionally, delivery was at 36–38 weeks to avoid stillbirth; but with close supervision pregnancies may go nearer to (but not beyond) term. *In labour:* Avoid acidosis and monitor the fetus (p 116). Avoid maternal hyperglycaemia (causes fetal hypoglycaemia). Monitor glucose; prevent hyperglycaemia with extra insulin (may need 5u/h) if β-sympathomimetics or glucocorticoids are used in preterm labour. Aim for vaginal delivery with a labour of <12h. Beware shoulder dystocia with macrosomic babies. With elective delivery, give normal insulin the evening before induction. During labour give 1L of 5–10% glucose/8h ivi with 1–2u insulin/h via a pump. Aim for a blood glucose of 4.5–5.5 mmol/L (check hourly). Insulin needs fall in labour and immediately postpartum. Halve insulin infusion rate post-delivery until a good meal is eaten; then return to pre-pregnancy regimen. Do a caesarean section if labour is prolonged.

Postnatal • Oral hypoglycaemics are contraindicated if breast feeding. • Encourage pre-pregnancy counselling *before* next pregnancy (p 94) to transfer to insulin. • Do a postpartum glucose tolerance test at 6 weeks.

Gestational DM (OGTT glucose ≥ 7.8, *OHCM* p 283) Incidence: 3%. medline:10563364 30% get full DM, so give lifelong dietary advice & follow-up. medline:3888735

Equations exist for giving risk of post-pregnancy DM from pre-pregnancy BMI (p 471), fasting plasma glucose, and months since delivery. medline 8475959 Other risk factors: age >30yrs; mothers who themselves have had low birth weights/IUGR (p 104); 1° relative with DM; unexplained stillbirth; medline 2408184 gestational DM before 27 weeks (or needing insulin). 2002,n 1636
medline 11967502 & 11852376

► Exercise, a good diet, and no smoking all help lower this risk.

Thyroid disease in pregnancy

(see also p 79)

► Whenever a mother isn't quite right postpartum, check her TSH & free T₄—but note that any apparent hypothyroidism may be transitory.

Biochemical changes in normal pregnancy NB: normal pregnancy mimics hyperthyroidism (pulse ↑, warm moist skin, slight goitre, anxiety).

► Thyroid binding globulin & T₄ output rise to maintain free T₄ levels, p 79.

► High levels of hCG (p 78) mimic thyroid stimulating hormone (TSH).

► There is reduced availability of iodine (in iodine limited localities).

► TSH may fall below normal in the first trimester (suppressed by hCG).

► The best thyroid tests in pregnancy are free T₄, free T₃, and TSH.

Pre-pregnancy hyperthyroidism Treatment options include antithyroid drugs (but 60% relapse on stopping treatment), radioactive iodine contraindicated in pregnancy or breast feeding: avoid pregnancy for 4 months after use), or surgery. Fertility is reduced by hyperthyroidism.

Hyperthyroidism in pregnancy (Usually Graves' disease.) There is ↑ risk of prematurity, fetal loss, and, possibly, malformations. Severity of hyperthyroidism often falls during pregnancy. Transient exacerbations may occur (1st trimester and postpartum). Treat with carbimazole or propylthiouracil (may cross placenta less). Keep dose as low as possible (reports of neonatal scalp defects are unreplicated in systematic studies, and antithyroid drugs are said to be relatively safe). Once under control, keep dose at ≤10mg/24h po, keeping T₄ at the top of the normal range. Monitor at least monthly. Some advocate stopping antithyroid drugs in the last month of pregnancy. Propylthiouracil is preferred postpartum (less concentrated in breast milk). If hyperthyroidism cannot be controlled by drugs, partial thyroidectomy can be done in the second trimester. Screen all women with a history of Hashimoto's or Graves' disease for TRAb (TSH-receptor stimulating antibodies); high levels can cause fetal and neonatal hyperthyroidism: blocking them causes hypothyroidism.

Hypothyroidism Relatively rare, it is associated with relative infertility. In untreated hypothyroidism there are increased rates of miscarriage, stillbirth, premature labour, and abnormality. T₄ requirements may increase up to 50%. Monitor adequate replacement by T₄ and TSH measurements each trimester. Requirements return to pre-pregnancy levels postpartum.

Postpartum thyroiditis Prevalence: 5%. Hyperthyroidism is followed by hypothyroidism (~4 months postpartum). 40% have permanent thyroid failure and 25% of those with transient disease become hypothyroid 4yrs later. TRAb antibodies are absent. The hyperthyroid phase does not usually need treatment as it is self-limiting. If treatment is required β-blockers are usually sufficient. Antithyroid drugs are ineffective because thyrotoxicosis results from thyroid destruction causing increased thyroxine release, rather than increased synthesis. The hypothyroid phase is monitored for 6 months, and the woman treated if she becomes symptomatic. Withdraw treatment after 6–12 months for 4 weeks to see if long-term therapy is required. Hypothyroidism may be associated with postpartum depression, so check thyroid status of women with postpartum depression.

Neonatal thyrotoxicosis Seen in 1% of babies of women with past Graves' disease, due to TSH-receptor stimulating antibodies crossing the placenta. Signs: fetal tachycardia (>160/min) in late pregnancy ± intrauterine growth restriction. If mother has been on antithyroid drugs signs may not be manifest until the baby has metabolized the drug (7–10 days postpartum). Test thyroid function in affected babies frequently. Antithyroid drugs may be needed. It resolves spontaneously at 2–3 months, but perceptual motor difficulties, and hyperactivity can occur later in childhood.

Jaundice in pregnancy

►Get expert help *promptly*. Jaundice in pregnancy may be lethal. Know exactly what drugs were taken and when (*prescribed* or *over-the-counter*). Where has she travelled to? What life-style or occupational risks are there?

Jaundice occurs in 1 in 1500 pregnancies. Viral hepatitis and gallstones may cause jaundice in pregnancy and investigation is similar in the non-pregnant. Those with Gilbert's and Dubin–Johnson syndrome (*OHCM* p 722–6) do well in pregnancy (jaundice may be exacerbated with the latter).

Intrahepatic cholestasis of pregnancy¹ Incidence 0.1–1.5% pregnancies in Europe, 9–15% S America. There is pruritus, especially of palms and soles in the second half of pregnancy. Liver transaminases are mildly raised (<300u/L) in 60%, raised bilirubin in 25%. Exclude viral hepatitis. There is risk of preterm labour (60%), fetal distress (33%), and stillbirth (2%), so monitor fetal wellbeing; aim for delivery by 38 weeks. Give vitamin K to the baby at birth. Ursodeoxycholic acid reduces pruritus and abnormal LFTs. Symptoms resolve within days of delivery. It is a contraindication to oestrogen-containing contraceptive pills and recurs in 40% pregnancies.

Acute fatty liver of pregnancy Incidence: 1 : 9000–13,000 deliveries—so it is rare but grave. The mother develops abdominal pain, jaundice, headache, vomiting, ± thrombocytopenia, and pancreatitis. There is associated pre-eclampsia in 30–60%. It usually occurs after 30 weeks. There is hepatic steatosis with microdroplets of fat in liver cells. Deep jaundice, uraemia, severe hypoglycaemia and clotting disorder may develop causing coma and death. Give supportive treatment for liver and renal failure and treat hypoglycaemia vigorously (CVF line). Correct clotting disorder with vitamin K, fresh frozen plasma, and antithrombin III. Expedite delivery. Epidural anaesthesia is contraindicated. Beware PPH and neonatal hypoglycaemia. Mortality can be as low as 18% maternal and 23% fetal. Other causes of fatty liver (steatosis): yellow fever, tetracycline, sodium valproate, Reye's syndrome (p 756).

Some other causes of jaundice in pregnancy

- Viral hepatitis; ALT, eg >200u/L; maternal mortality ↑ in E virus, *OHCM* p 578.
- The jaundice of severe pre-eclampsia (hepatic rupture and infarction can occur); ALT <500u/L; bilirubin <86µmol/L.
- Rarely complicating hyperemesis gravidarum (can be fatal); ALT <200u/L.
- Hepatitis may occur if halothane is used for anaesthesia (so avoid it).
- HELLP syndrome (haemolysis, elevated liver enzymes, and low platelet count). Incidence in pregnancy: 0.1–0.6%; in pre-eclampsia: 4–12%. It causes: upper abdominal pain, malaise, vomiting, headache, jaundice, microangiopathic haemolytic anaemia, DIC, LDH ↑, ALT ↑ <500u/L, bilirubin <86µmol/L. It recurs in 20%. Treatment: get expert help. Admit; deliver if severe.

Hepatitis B Check HBsAg in all women with jaundice. Staff should always be careful to avoid contact with blood during delivery (esp. if HBsAg +ve) and be especially careful with disposal of 'sharps'. Give 200u specific hepatitis B immune globulin (HBIG) IM to the anterolateral thigh to babies of affected mothers at birth—*plus* hepatitis B vaccine (0.5mL EngerixB® into the other anterolateral thigh) at birth, 1 month and 6 months. Offer vaccination to *all* the family. Do serology at 12–15 months old. If HBsAg –ve and anti-HBs is present, the child has been protected.

Tests Do all the usual tests (*OHCM* p 206), eg urine tests for bile, serology, LFTs, and ultrasound. Special tests may be needed to show fat, eg oil red O, to fresh-frozen biopsy samples—if coagulation status allows biopsy.

Malaria in pregnancy

In any woman who presents with odd behaviour, fever, jaundice, sweating, DIC, fetal distress, premature labour, seizures, or loss of consciousness, always ask yourself: ► *Could this be malaria?* If so, do thick and thin films. Confirm (or exclude) pregnancy. Seek expert help (eg from Liverpool, below).

Falciparum malaria can be particularly dangerous (and complicated) in pregnancy, particularly in those with no malaria immunity. In some places, such as Thailand, malaria is the leading cause of maternal mortality. Cerebral malaria has a 50% mortality in pregnancy. Third stage placental autotransfusion may lead to fatal pulmonary oedema. Hypoglycaemia may be a feature of malaria in pregnancy (irrespective of whether quinine has been given).¹

Other associations between *falciparum* malaria and pregnancy are anaemia, abortion, stillbirth, low birth weight, and prematurity. PPH is also more common. Hyperreactive malaria splenomegaly (occurs typically where malaria is holoendemic) may contribute to anaemia via increased haemolysis.

Vivax malaria is less dangerous, but can cause anaemia and low birth weight.

Treatment of malaria is as described in *OHCM* p 564. Chloroquine and quinine (in full dosage) appear to be safe in pregnancy, but beware hypoglycaemia. For *falciparum* malaria, expect to use quinine as chloroquine resistance is widespread. Mefloquine can be used in the second and third trimesters for the treatment of uncomplicated *falciparum* malaria.

Transfer earlier rather than later to ITU. Women with a haematocrit <20% should get a slow transfusion of packed cells, if compatible, fresh, pathogen-free blood is available. Remember to include the volume of packed cells in calculations of fluid balance. Exchange transfusions may be needed—if facilities are available.

During labour, anticipate fetal distress, fluid-balance problems, and hypoglycaemia. Monitor appropriately. Send placenta for microscopy for malarial parasites. If they are present the baby may be infected (the parasites can cross the placenta—especially in non-immune mothers) and should be treated.²

Prevention in UK women Advise against visiting malarious areas. If is unavoidable, give prophylaxis (*OHCM* p 564). Emphasize importance of preventive measures such as mosquito nets and insect repellents. Chloroquine and proguanil are used in normal doses if *P falciparum* strains are sensitive—but with proguanil, concurrent folate supplements should be given.

Maloprim® (dapsone+pyrimethamine) is contraindicated in first trimester; if given in second or third trimesters, give folate supplements too.

Mefloquine should be avoided as prophylaxis (exclude pregnancy before use, and avoid pregnancy for 3 months after use).

Trials of chemoprophylaxis in mothers living in endemic areas

These show that chemoprophylaxis improves birth weight (↑by ~250g—and there are fewer very low birth weight babies). Red cell mass also improves. The effect of universal maternal chemoprophylaxis on the emergence of drug-resistant strains of malaria has not been quantified—and neither have sophisticated outcome studies been done on general neonatal wellbeing with maternal prophylaxis. Coarse analysis of the figures suggests that there is no adverse effect on either maternal or fetal mortality.³ In some studies primips benefit more from chloroquine than multip,⁴ even though the area (Cameroon) is officially chloroquine-resistant.

► If in doubt, phone an expert, eg, in the UK, at Liverpool (0151 708 9393).

¹ WHO 2001 *Management of Severe & Complicated Malaria* ² R Hurley 1995 in *Medical disorders in Obs Pract* 3e Blackwell 562 ³ P Garner 1997 *E-BM* 2 15 ⁴ *Tropical Med & Internat Health* 2002 7 29 [011851952] & M Motta 2002 *Lancet* 359 524 [011853823]

Renal disease in pregnancy

Note: Values considered normal when not pregnant may reflect decreased renal function in pregnancy. Creatinine $>75\mu\text{mol/L}$ and urea $>4.5\text{mmol/L}$ merit further investigation. See p 79. Glycosuria in pregnancy may reflect altered renal physiology and not necessarily imply hyperglycaemia.

► Treat asymptomatic bacteriuria in pregnancy. Check that infection and bacteriuria clear with treatment.

Asymptomatic bacteriuria This is found in 2% of sexually active women, and is more common (up to 7%) during pregnancy. With the dilatation of the calyces and ureters that occurs in pregnancy, 25% will go on to develop pyelonephritis, which can cause fetal growth restriction, fetal death, and premature labour. This is the argument for screening all women for bacteriuria at booking. If present on 2 MSUs treatment is given (eg amoxicillin 250mg/8h po with a high fluid intake). Test for cure after 1 and 2 weeks. If the organism is not sensitive to amoxicillin, consider nitrofurantoin 50mg/6h po with food.

Pyelonephritis This may present as malaise with urinary frequency or as a more florid picture with raised temperature, tachycardia, vomiting, and loin pain. It is common at around 20 weeks and in the puerperium. Urinary infections should always be carefully excluded in those with hyperemesis gravidarum and in those admitted with premature labour. Treatment is with bed rest and plenty of fluids. After blood and urine culture give iv antibiotics (eg ampicillin 500mg/6h iv, according to sensitivities) if oral drugs cannot be used (eg if vomiting). Treat for 2–3 weeks. MSUs should be checked every fortnight for the rest of the pregnancy. 20% of women having pyelonephritis in pregnancy have underlying renal tract abnormalities and an ivu or ultrasound at 12 weeks' postpartum should be considered. In those who suffer repeated infection, nitrofurantoin (100mg/24h po with food) may prevent recurrences. Avoid if the glomerular filtration rate is $<50\text{mL/min}$. SE: vomiting, peripheral neuropathy, pulmonary infiltration, and liver damage.

Chronic renal disease With mild renal impairment without hypertension there is little evidence that pregnancy accelerates renal disorders. Patients with marked anaemia, hypertension, retinopathy, or heavy proteinuria should avoid pregnancy as further deterioration in renal function may be expected and fetal loss is considerable (up to 60%). Close collaboration between physicians and obstetricians during pregnancy in those with renal disease is the aim. Induction of labour may become advisable in those with hypertension and proteinuria, or if fetal growth is retarded.

Pregnancy for those on dialysis is fraught with problems (fluid overload, hypertension, pre-eclampsia, polyhydramnios). A 50% increase in dialysis is needed. Live birth outcome is 50%, at best. Outcome is better for those with renal transplants.

Obstetric causes of acute tubular necrosis Acute tubular necrosis may be a complication of any of the following situations:

- Septicaemia (eg from septic abortion or pyelonephritis).
- Haemolysis (eg sickling crisis, malaria).
- Hypovolaemia, eg in pre-eclampsia; haemorrhage (APH, eg abruptio, PPH, or intrapartum); DIC; abortion—or adrenal failure in those on steroids not receiving booster doses to cover labour.

Whenever these situations occur, monitor urine output carefully (catheterize the bladder). Aim for $>30\text{mL/h}$ output. Monitor renal function (U&E, creatinine). Dialysis may be needed (OHCM p 266).

Epilepsy in pregnancy

► They key to successful pregnancy is access to good preconception counselling (p 94). ► If seizures occur in pregnancy, think *could this be eclampsia?* Epilepsy *de novo* is rare in pregnancy. Epilepsy affects ~0.5% of women of childbearing age so a unit with 3000 deliveries per year has ~15 pregnant epileptic women at any one time. Seizure rate during pregnancy increases in ~25% of epileptic women, reduces in ~22% and is unchanged in the remainder. Where seizure rate increases this is often associated with lack of compliance with medication, or sleep deprivation. It is unusual for seizures to recur in pregnancy when preceded by a long seizure-free period.

Complications—*Maternal*: ↑Risk of 3rd trimester vaginal bleeding, 1% convulse in labour. *Fetal*: ↑Risk of: prematurity; stillbirth; neonatal and perinatal death; haemorrhagic disease. Epilepsy affects 3% of offspring when the mother has partial epilepsy (5–10% if she has idiopathic generalized epilepsy).

In a recent retrospective population-based study from Scotland,¹ neonatal withdrawal was seen in 20% of those exposed to antiepileptics. Congenital malformations occurred in 14% of exposed pregnancies, compared with 5% of non-exposed sibs, and developmental delay in 24% of exposed children, compared with 11% of non-exposed sibs. 31% of exposed children had either major malformation or developmental delay, 52% of exposed children had facial dysmorphism compared with 25% of those not exposed, 31% of exposed children had childhood medical problems (vs 13% of non-exposed sibs), and 20% had behaviour disorders (vs 5% of non-exposed sibs). *Cleft lip*: Epilepsy in the mother (but not father) is associated with this. Relative risk for a fetus having clefts compared to the non-epileptic population is 1.0 if the mother develops epilepsy after the pregnancy; 2.4 if she develops it after conception (but has no drugs); and 4.7 if the fetus is exposed to anticonvulsants.

Malformation is commoner if ≥2 anticonvulsants are used, or high doses, and if a sibling is already malformed. *Fetal valproate syndrome* (10% of those born to mothers having valproate regularly in pregnancy.) Signs: major organ system anomalies ± autism ± small ears, small broad nose, flat philtrum, a long upper lip, shallow philtrum & micro/retrognathia.² Neural tube defects are commoner with valproate (& carbamazepine) so screen for these (p 166).

Phenytoin and phenobarbital cause congenital heart disease and cleft lip.

Management ► Get expert help. Avoid trimethadione and paramethadione (both very teratogenic). Where anticonvulsants are needed the dose of the chosen drug should be kept as low as possible. Aim for 1 drug only. Be careful interpreting plasma phenytoin levels as free levels may remain the same despite a reduced bound level. Give folic acid supplements, eg 5mg/24h from prior to conception and vitamin K₁ 20mg/24h to the mother from 36 weeks if she is taking phenytoin, primidone, or phenobarbitone. Treat status epilepticus as in the non-pregnant except that the fetus should be monitored. It is associated with significant fetal and maternal mortality. Deliver in hospital (1–2% epileptic women convulse in labour and the subsequent 48 hours). Give vitamin K to the baby at birth (p 244).

Neonates can suffer barbiturate withdrawal with hyperexcitability, tremulousness, impaired suckling and, occasionally, seizures at ~1 week if not breastfed. Usually, babies only require supervision, but if very tremulous, or if there are seizures, they may be given 3–5mg/kg/day phenobarbital.

Review drugs postnatally. There may be ↑susceptibility to fits while bathing. Mothers may breast feed (phenobarbital can cause drowsiness in the baby).

1 J Dean 2002 *J Med Genetics* 39 251 [11950853] 2 D Renier 2001 *J Neurosurg* 95 78 [11702867] 62% had musculoskeletal abnormalities, 26% had cardiovascular abnormalities, 22% had genital abnormalities, and 16% had pulmonary abnormalities. [11223853]

Connective tissue diseases in pregnancy

Rheumatoid arthritis is usually alleviated by pregnancy (but exacerbations may occur in the puerperium). Methotrexate use is contraindicated (teratogenic); sulfasalazine may be used (give extra folate). Azathioprine use may cause intrauterine growth restriction and penicillamine may weaken fetal collagen. Non-steroidal anti-inflammatories can be used in the first and second trimesters but are not recommended in the third as they can cause premature closure of the ductus arteriosus and late in pregnancy have been associated with renal impairment in the newborn. Congenital heart block is a rare fetal feature. Deliver babies with heart block by caesarean section.

Systemic lupus erythematosus SLE exacerbations are commoner in pregnancy and the puerperium. Most are mild to moderate involving skin, but those with renal involvement and hypertension may deteriorate and are prone to pre-eclampsia. Of those with SLE glomerulonephritis and a creatinine $>130\mu\text{mol}$ prior to conception only 50% achieve a live birth. Pre-eclampsia, oligohydramnios, and intrauterine growth restriction may occur. Both hydralazine and methyldopa can be used in pre-eclampsia.

Planned pregnancy should be embarked on after 6 months stable disease without requiring cytotoxic suppression. Disease suppression may be maintained with azathioprine and hydroxychloroquine. Aspirin 75mg daily should be started prior to conception and continued throughout pregnancy and the fetus should be carefully monitored.

Rarely the fetus is affected by passively acquired maternal anti-Ro antibodies that can cause a self-limiting sunlight sensitive rash (usually face & scalp) for which no treatment is required, but more rarely can cause irreversible damage to the fetal heart conduction system causing congenital heart block (~65% of these neonates require a pacemaker). Deliver as above.

Mothers requiring $\geq 7.5\text{mg}$ daily prednisolone in the 2 weeks before delivery should receive hydrocortisone 100mg/6h iv in labour.

Antiphospholipid syndrome Those affected have antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies) \pm past arterial thrombosis, venous thrombosis, or recurrent pregnancy loss. It may be primary, or follow other connective tissue disorder (usually SLE in which it occurs in 10%). *Outcome:* Untreated, $<20\%$ of pregnancies proceed to a live birth due to 1st trimester loss or placental thrombosis (causes placental insufficiency, leading to intrauterine growth restriction and fetal death).

Careful regular fetal assessment (Doppler flow studies and ultrasound for growth are required from 20 weeks) as appropriate obstetric intervention can substantially increase the number of live-born babies.

Management: Affected women are treated from conception with aspirin 75mg daily (heparin sc/12h until 34 weeks improves outcome). Those who have suffered prior thromboses receive heparin. See p 151.

Postpartum use either heparin or warfarin (breastfeeding contraindicated with neither) as risk of thrombosis is high.

Pregnancies in those with SLE (especially with renal disease) and antiphospholipid syndrome require specialist management.

Maternal mortality

... beliefs, art, work—none of these are worth sacrificing your life for, the on
... worth sacrificing life for is life itself. All mothers sacrifice themselves here
... agree some pay the highest price, and it is to them that this page is dedicate

Maternal mortality is defined in the UK as the death of a mother while pregnant or within 42 days of termination of pregnancy, from any cause related to or aggravated by the pregnancy or its management, but not from accidental or incidental causes. Deaths are subdivided into those from 'direct causes'—those in which the cause of death is directly attributable to pregnancy (eg abortion, eclampsia, haemorrhage)—and indirect deaths—those resulting from previous existing disease or disease developed during pregnancy, and which were not due to direct obstetric causes but were aggravated by pregnancy (eg heart disease). Late deaths are those occurring between 42 days and 1 year after termination, miscarriage, or delivery that are due to direct or indirect maternal causes.

History of a successful preventive enterprise Since 1952 there have been 3-yearly confidential enquiries into maternal deaths. Prior to 1979, as many deaths were considered to have had 'avoidable factors' (this term was used to denote departures from acceptable standards of care by individuals, including patients) but since 1979 the wider term of 'substandard care' has been used to cover failures in clinical care and other factors, such as shortage of resources and back-up facilities.

Maternal mortality has almost halved every decade since reports have been issued (deaths per 100,000 maternities have been 67.1 in 1955–7, 33.3 in 1964–6, 11 in 1973–5, and 11.4 in 1997–9 of which direct deaths were 5 per 100,000).¹ Rates are lowest for women aged 20–24 years, rising markedly in the over-35s, and for first pregnancies (rising with increasing parity). It has been estimated that maternal death rates/100,000 births are 640 in Africa; 420 in Asia; 270 in Latin America; 30 in all developed countries and < 10 in Northern and Central Europe.

In 1997–9, 378 deaths were recorded including 107 late deaths. Of these 106 were direct obstetric deaths; 136 were indirect obstetric deaths and 29 were coincidental (in no way related to pregnancy, eg car accident). There was no strong association with geographical region. Risk is increased for non-white women, especially those from India, Bangladesh, and Pakistan,¹ (2x more likely to die); those socially disadvantaged (20x more likely); travellers; those suffering domestic violence; the obese (body mass index >35 at booking; see p 471); those aged <18 yrs; and those undergoing *in vitro* fertilization.

In 1997–9 thromboembolism was the chief cause of direct death in the UK (33% of deaths). Other direct causes: early pregnancy (16%) (mainly ectopic pregnancy at 12.3%); hypertensive disorders (14.1%); genital tract sepsis (13.2%); amniotic fluid embolism (7.5%); haemorrhage (6.6%); fatty liver of pregnancy (3.7%), anaesthetic deaths (2.8%). 34% died before delivery.

The death rate from caesarean section for the 1985–90 period was 0.33 per 1000 operations. Mortality is increased 5-fold (vs vaginal delivery), for all sections—and 12-fold for emergency sections.¹

The maternal mortality rate was similar to the previous triennium for direct deaths, and for the first time there were more indirect than direct deaths. Care was considered substandard in 60.4% of cases of direct death (in a major way in 50%) in 1997–9, ('substandardness' includes pregnant women who refuse medical advice).

1 2001 Why Mothers Die The Confidential Enquiries into Maternal Deaths in the United Kingdom 1997–9. RCOG **2** DoH 1994 Report on Confidential Enquiries into Maternal Deaths in the United Kingdom 1988–90, London, HMSO

Perinatal mortality

This is the number of stillbirths and deaths in the first week of life/1000 total births. Stillbirths only include those fetuses of >24 weeks' gestation, but if a fetus of <24 weeks' gestation is born and shows signs of life, and then dies, it is counted as a perinatal death in the UK (if dying within the first 7 days). Neonatal deaths are those infants dying up to and including the 28th day after birth. Other countries use different criteria—including stillbirths from 20 weeks and neonatal deaths up to 28 days after birth, so it is not always easy to compare statistics.

Perinatal mortality is affected by many factors. Rates are high for *small* (61% of deaths are in babies <2500g) and *preterm* babies (70% of deaths occur in the 5% who are preterm). See p 102 & p 180. *Regional variation* in the UK is quite marked with rates for the Oxford region being considerably lower than for Merseyside. There is a *social class variation* with rates being less for social classes 1 and 2 than for classes 4 and 5. *Teenage mothers* have higher rates than mothers aged 20–29. From 30yrs rates rise until they are 3-fold higher than the low-risk group (20–29 years) by the age of 40–45. *Second babies* have the lowest mortality rates. Mortality rates are doubled for fourth and fifth children, trebled by sixth and seventh (this effect is not independent of social class as more lower social class women have many children). Rates are lower for *singleton births* than for multiple. Rates are higher for the offspring of mothers of *Bangladeshi* or *West Indian* extraction living in the UK, but are the same for those of Pakistani and UK extraction.

Perinatal mortality rates in the UK have fallen over the years from rates of 62.5/1000 in 1930–5 to 7.8/1000 in the 1990s. Declining mortality reflects improvement in standards of living, improved maternal health, and declining parity, as well as improvements in medical care. The main causes of death are congenital abnormalities (22%), unclassified hypoxia (18%), placental conditions (16%), birth problems including cord problems (11%), and maternal conditions and toxemia (8%).

Of neonatal deaths the main cause is prematurity (49%); infection counting for 7%.

Examples of how changed medical care may reduce mortality:

- Worldwide, treatment of syphilis, antitetanus vaccination (of mother during pregnancy), and clean delivery (especially cord techniques), have the greatest influence in reducing perinatal mortality.¹
- Antenatal detection and termination of malformed fetuses.
- Reduction of mid-cavity procedures and vaginal breech delivery.
- Detection of placenta praevia antenatally.
- Prevention of rhesus incompatibility.
- Preventing progression of preterm labour.
- Better control of diabetes mellitus in affected mothers.
- Antenatal monitoring of 'at risk' pregnancies.

While we must try to reduce morbidity and mortality still further, this must not blind us to other problems that remain, such as the 'over-medicalization' of birth; the problem of reconciling maternal wishes to be in charge of her own delivery with the immediate needs of the baby; and the problem of explaining risks and benefits in terms that both parents understand, so that they can join in the decision-making process.

Prenatal diagnosis

'The first half of pregnancy can become a time of constant 'exams' to see if the baby can be allowed to graduate to the second half of pregnancy'.¹ Parents at high and, increasingly, those at low risk of having an abnormal baby are offered prenatal diagnosis if this might permit more effective treatment of the expected defect, or (more often) if they wish to terminate any abnormal pregnancy.

High-risk pregnancies • Maternal age >35 (chromosome defects).

• Previous abnormal baby or family history of inherited condition.

Problems ► Anxiety while false +ve results are sorted out is a big problem.

• Terminating normal fetuses, eg ♂ fetus of carriers of X-linked conditions.

• Most abnormalities are in low-risk groups (∴ missable by selective screening).

• Services available, their quality, and populations made eligible, vary widely.

• Termination of female fetuses in cultures valuing males more highly.

• Devaluation of positive view of handicapped or 'special needs' children.

Ultrasound At 11–14, then ~18 weeks, skilled operators can detect many external and internal structural anomalies. See p 118, and opposite.

Ultrasound is best at detecting externally impinging structural abnormalities, eg anencephaly/spina bifida. Internal structural abnormality detection rate, eg for heart disease and diaphragmatic hernia, remains <50%. Fetuses with false +ve suggestion of abnormality are mostly associated with 'soft signs' on ultrasound, eg nuchal thickening (eg trisomy 21), choroid plexus cysts (trisomies 18 and 21), and echogenic bowel (trisomy 21 and cystic fibrosis). In one large study ($n=33,376$ fetuses; 725 abnormal; 55% of which detected prenatally) use of ultrasound 'soft signs' ↑total detection rate from 51% to 55% but ↑false +ves 12-fold from one in 2332 to one in 188.²

α-Fetoprotein (AFP) AFP is a glycoprotein synthesized by the fetal liver and GI tract. Fetal levels fall after 13 weeks, but maternal (transplacental) serum AFP continues to rise to 30 weeks. Maternal AFP is measured at 17 weeks. In 10% with a high AFP there is a fetal malformation, eg an open neural tube defect (but closed defects are missed), exomphalos, posterior urethral valves, nephrosis, GI obstruction, teratomas, Turner's syndrome (or normal twins). In ~30% of those with no malformation, there is an adverse outcome, eg placental abruption and third trimester deaths. ► Monitor closely. 1 in 40 with a low AFP have a chromosomal abnormality (eg Down's). AFP is lower in diabetic mothers. NB: as this test is nonspecific on its own, it is of use for preliminary screening; those with abnormal values may be offered further tests (see below, and p 168 for the 'triple test').

Amniocentesis (p 120) is carried out under ultrasound guidance. Fetal loss rate is 0.5–1% at ~16 weeks' gestation, but ~5% for early amniocentesis at 10–13 weeks. Amniotic fluid AFP is measured (a more accurate screen for neural tube defects than maternal serum), and cells in the fluid are cultured for karyotyping and enzyme and gene probe analysis. Cell culture takes 3 weeks, so an abnormal pregnancy must be terminated at a late stage.

Chorionic villus biopsy At 10+ weeks, placenta is sampled by transcervical catheter or transabdominal needle under ultrasound guidance. Karyotyping takes 2 days, enzyme and gene probe analysis 3 weeks, so termination for abnormality is earlier, safer, and less distressing than after amniocentesis. Fetal loss rate is ~4%. Use up to 20 weeks (cordocentesis preferable thereafter).³ It does not detect neural tube defects—and may cause fetal malformation.

Fetoscopy is carried out at ~18wks with ultrasound guidance to find external malformations, do fetal blood samples, or biopsy. Fetal loss rate is ~4%.

High resolution ultrasound and fetal nuchal translucency

- Early scans (at 11–14 weeks) may detect 59% of those with structural abnormality and 78% of those with chromosome abnormality. It is especially good for detecting CNS defects, neck abnormalities, GI, and renal defects: less good for spina bifida, heart, and limb defects. With a combination of early and later scans up to 81% of malformations may be diagnosed.⁴
- Fluid accumulation in the neck at 10–14 weeks' gestation (increased fetal nuchal translucency, FNT) reflects fetal heart failure, and is typically seen in any serious anomaly of the heart and great arteries.^{5,6}
- There is a strong association between chromosomal abnormality and FNT. In one study, 84% of karyotypically proven trisomy 21 fetuses had a nuchal translucency >3mm at 10–13 weeks' gestation (as did 4.5% of chromosomally normal fetuses).
- The greater the extent of FNT, the greater the risk of abnormality.
- Nuchal translucency screening may be used to see who may benefit from more invasive chorionic villus sampling (or amniocentesis, which may delineate the precise chromosomal abnormality (eg trisomies). Note: positive predictive value of screening is 4% so 96% of women with a 'positive' test undergo an 'unnecessary' invasive procedure (chorionic villus sampling in the first trimester or amniocentesis in the second trimester). If nuchal screening was used as the *only* screening test 2 or 3 normal pregnancies would be lost after chorionic villus sampling, and 1 after amniocentesis, for every 4 pregnancies correctly detected with trisomy 21.
- It is useful for screening twins as early detection is best, for if selective fetocide is to be used risk of miscarriage is 3-fold higher if done after 16 weeks. Monochorionic twins have a higher false +ve rate for nuchal translucency thickness than dichorionic twins or singletons.
- Note that the degree of neck flexion during the ultrasound examination may influence nuchal measurements.
- Note adding nasal bone screening at 11–14 week ultrasound may increase sensitivity and reduce the false positive rate of screening for Down's syndrome.⁷

Blood tests to detect Down's syndrome

The triple test Maternal serum AFP, unconjugated (o)estriol and total human chorionic gonadotrophin (HCG) are assessed in relation to maternal age, weight, and gestation (determined by ultrasound) to estimate risk of Down's syndrome (and, in some places, of neural tube defects). See also screening by fetal nuchal translucency ultrasound (p 167).

Problems with the triple blood test—the triple conundrum

1 It may not be the best test: Some authorities say that adding unconjugated estriol to the test could merely increase uncertainty. Also, there is evidence that free HCG is a better marker than total HCG. Quadruple tests measure inhibin-A. *Integrated tests* becoming available in the UK measure pregnancy-associated protein-A (PAP-A) and nuchal thickness from 3 measurements in the first trimester, and measurements of blood α -feto protein, unconjugated oestriol, HCG, inhibin-A taken 2 to 4 weeks later. These should detect 85% of affected pregnancies with a false positive rate of 1%.¹ As the age at which mothers have their babies increases, any relative benefit of the triple test over age-related screening diminishes.

2 The cost-benefit sums are hard to calculate.

3 The emotional cost to the mother is impossible to calculate: From the parents' point of view, a telling statistic is that 56 out of every 57 women under 37yrs old who had a +ve test, proved, after amniocentesis, *not* to have an affected fetus. Amniocentesis causes fetal loss, and these losses will almost always be of normal babies. Newer screening regimens in the first trimester will go some way to mitigating distress and anxiety. One *stop assessment* offers nuchal screening and rapid result analysis of β HCG and PAP-A between 10 weeks 3 days and 13 weeks 6 days. It can achieve detection rates of 95% of all aneuploidies, 86% trisomy-21, 100% of trisomy-18 and trisomy-13. In one study ($n=4190$), 97.6% took up screening; false positive screening occurred in 6.7%: 7 cases had neural tube defects, 2 ventral wall defects were picked up on scan. 200 accepted chorionic villus sampling: there was fetal demise in 2 in next 28 days 1 of which had Down's.¹

We have no idea of the best way of counselling parents before the test. If you just hand out a leaflet, few will read it, and then when it comes to amniocentesis and termination, many will refuse—and the screening test wastes money, as well as laying health authorities open to litigation: 'I never understood that I might lose a normal baby ...' The alternative is to provide full details at the time of the initial blood test. The irony is that gaining informed consent is then the most expensive part of the test, and one which itself could cause much distress. Imagine an overjoyed expectant mother arriving in the clinic serenely happy in fulfilling her reproductive potential: the quintessence of health. She leaves only after being handed ethical conundrums of quite staggering proportions, involving death, disease, and human sacrifices, and a timetable for their resolution that would leave even the most fast-moving philosopher breathless and disorientated, and which may leave her forever bereft of one of Nature's most generous gifts: the fundamental belief in one's own wholeness.

Example of counselling before doing a less than perfect test, without distressing women with talk of having terminations for what may be a very much wanted pregnancy.

'We've got a package of tests we often use to see how the pregnancy is going on—would you like us to get on with what seems best (nothing is perfect!)—or are you the sort of person who would want to know in detail about each test? If so, we can arrange a time for you to come back for further discussions.'

This allows the mother to pace her access to information in her own way—and each person's needs in this regard will be different.

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who are you, little i
(5 or 6 years old)
peering from some high window... e e cummings

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Introduction

Manufacturing organs is the point of embryology...

Growth and development is the essence of childhood...

Protection from a cynical world is the *raison d'être* of the family...

And the rôle of paediatrics is to align the above in the most constructive way. This is different for each individual, and different for each society, but forever the same in striving to allow each child to fulfil his or her own potential. But what *is* this potential? ► **Potential is only ever lost**; as with childhood itself, it slips through our fingers while we look the other way. The egg has more potential than the embryo (its sex, for example, may not yet be determined), and the child has more potential than the medical student who is perpetually cutting off lines of enquiry, and specializing in one branch of biology.

So with this in mind we must redefine paediatrics as the science which enables children to lose their potential **slowly**. 'My word!—how you've grown!' is the silliest expression paediatrics knows—what marks out the human infant from almost all others is the slowness of growth and development. Don't think of the adolescent growth spurt, think why development is typically so slow. What is all this time **for** (note that we do not grow so fast during the adolescent growth spurt as we do in early life). Why does the growth rate slow in middle years of childhood? Perhaps because this gives us time to learn to control the most dangerous piece of equipment we will ever lay our hands on—the human brain.

Be they ever so sound in body and mind, children need to know (to ask themselves) whether to be happy or free—creative or reasonable. Here we reach the heart of paediatrics which is equipping the child to lead him or herself—letting the child decide, while we, humble paediatricians, forever run one step behind, pushing our trolley of wares down a one-way street.

The newcomer to paediatrics is presented with a double difficulty: not only are children, in many important respects, quite unlike adults, but the neonate is unlike the toddler—who is unlike the school child or the adolescent. A whole book along the lines of this work could be written on each of these groups of children, and still give a rather sketchy account. But the newcomer should know, in case he or she becomes downhearted, that no one can claim mastery of this subject. Rather than aiming for completeness, the aim is to be an effective paediatrician. This entails not endless hours in the library, but time on the wards (and in patients' homes, if the reader is lucky enough to have access to these) to enlarge on those innate skills of listening and talking to mothers and children in a language that each understands. Doctors who look after children have unrivalled opportunities to practise a holistic art, and it is possible that these doctors have a special skill here. We note with great interest that most patients between the ages of 15 and 20 who have acute leukaemia treated by paediatricians are cured—up to 63%, whereas only <50% of this group survive if treated in adult units.

Eliciting the history

There are 3 aims. **1** Reaching a diagnosis or differential diagnosis. **2** Placing the diagnosis in the context of the child and family. There is no one treatment for pneumonia, or diabetes, or anything else (same disease, but different contexts require different treatments). **3** To establish a good relationship with the parents and the child, so that if there is nothing the matter with the child the parents feel able to accept this. Conversely, if there is serious or untreatable illness, the aim is to build up trust, so that the parents are able to accept the best advice, and the child feels that he or she is in safe, friendly hands. So if possible, avoid any hurry or distractions. Introduce yourself; explain your rôle in the ward or consulting room. Take a history from as many sources as possible, the child, mother, father, and any significant other—but beware allowing the child to feel marginalized, so address yourself to the child first, and last.

Presenting complaints Record the child's and mother's own words.

The present illness When and how did it start? Was he/she well before? How did it develop? What aggravates or alleviates it? Has there been contact with infections? Has the child been overseas recently?

Especially in infants, enquire about feeding, excretion, alertness, and weight gain. After ascertaining the presenting complaint further questioning is to test the various hypotheses of differential diagnosis.

Past *In utero*: Any problems (eg PET, rubella, Rh disease); drug exposure?

At birth: Gestation, duration of labour, mode of delivery, birth weight, resuscitation required, birth injury, malformations.

As a neonate: Jaundice, fits, fevers, bleeding, feeding problems.

Ask about later illnesses, operations, accidents, screening tests, drugs allergies, immunization, travel, and drug or solvent abuse.

Development (p 290) Does the mother remember milestones reliably?

Drugs Prescribed, recreational, *in utero*, and over-the-counter. Drug intolerances, adverse drug reactions, and true allergies.

Family history Stillbirths, TB, diabetes mellitus, renal disease, seizures, jaundice, malformations, others. Are siblings and parents alive and well? Find out about late-onset diseases with a genetic component.*

Consanguinity is common in some cultures and may be relevant to disease.

Social history It may be vital to know who the father is, but damaging to ask the mother directly. Asking about the 'family unit', or drawing a family tree (OHCM p 33) may be one way forward. Be prepared to allow information to surface slowly, after chats with friendly nurses. Ask about play, eating, sleeping (excessively wrapped or liable to cold?), schooling and pets. Who looks after the child if the parents work? What work do they do? Ask about their hopes, fears, and expectations about the child's illness and his stay in hospital.

Privacy, dignity, and confidentiality These are easy to pay lip-service to, but very hard to ensure in busy paediatric wards, where space is at a premium. During an average stay in hospital most patients and their relatives will overhear confidential exchanges, and only a few will ever recall being offered a screen to preserve privacy during examinations.¹

► If the family does not speak your language, find an interpreter

*For example, if a parent has had a myocardial infarction before 40 years old, do serum lipids (>40% of these children will turn out to have hyperlipidaemia: the sooner it is treated, the better).

Systems review: examples of pertinent questions

	The neonate:	The toddler:	The older child:
Cardiorespiratory:	Tachypnoea; grunts Wheeze; cyanosis; cold sweats (heart failure)	Cough Exertional dyspnoea	Cough; wheeze Sputum; chest pain
Gastrointestinal:	Appetite p&v; feeding problems; Stool frequency; jaundice	Appetite p&v Stool frequency	Appetite p&v; abdominal pain Stool frequency
Genitourinary:	Wet nappies (how often?)	Wet nappies (how often?)	Haematuria; dysuria Sexual development
Neuromuscular:	Fits; odd attacks 'jitters' Feeding ability	Fits Drowsiness Hyperactive? Vision Hearing Gait	Headaches; Fits; Odd sensations; Drowsiness Academic ability Vision; Hearing Co-ordination
ENT and teeth:	Noisy breathing	Ear discharge Teeth eruption	Earache/discharge Sore throat

General questions: Growth, weight, failure to thrive, fatigue, lumps, itch, fevers, bleeding tendency, family interaction questions.

Physical examination: a method

No single routine will suffice for all children. If the child is very ill, examination must be limited (p 175). Furthermore, points in the examination assume varying importance depending on age (for neonates, see p 231). But it is helpful to have some sort of standard—on this page we look at a boy of 3 years who is being seen because of vomiting. Not everything on even this slimmed-down page will need to be done. The more experienced you are, the better you will be able to judge when and how to take short cuts. Do not mistake taking short cuts for being lazy. Use the time you save to be available to answer questions, and to address the fears and hopes of the parents and their child. NB: there is no correct order: be opportunistic, eg with younger children on a lap.

- 1 Wash and warm your hands. Encourage both parents to be present.
- 2 Regard the child (eg while feeding). Is he ill or well? Restless, still, or playing? If crying, is it high pitched or normal? Is he behaving normally? Any jaundice, cyanosis, rashes, anaemia, or dehydration (p 250)?
- 3 Talk to the child. Explain what you are doing. This helps you both relax, and enjoy the occasion (not a trivial point: a happy doctor is an engaged doctor, and an engaged doctor is amenable to receiving subtle signs and covert communications). If he seems anxious, examine his teddy in a playful way, to allow him to build up trust.
- 4 If the child is quiet or asleep, now is the time to get any listening done, so examine his heart, lungs, and abdomen. Start by hand-on, then move to using a warm stethoscope. Undress in stages.
- 5 Examine finger nails, then the hands, radial pulse, BP, axillary and neck nodes, neck lumps. Is there neck stiffness? (a 'useless' sign in infants)
- 6 Size and shape of head (p 294). Facial symmetry. Does the anterior fontanelle (between the parietal and frontal bones) feel tense (intracranial pressure ↑), or sunken (dehydration)?
- 7 Is there mucus in the nose? Leave ears and throat alone at this stage.
- 8 Count the respirations. Is there intercostal recession (inspiratory indrawing of the lower costal area, signifying ↑work of breathing?)
- 9 Percuss the chest if >2yrs old (not very reliable even then), and palpate the abdomen. Is it distended, eg by spleen, liver, fluid, flatus, faeces?
- 10 Undo the nappy, if worn. Have an MSU pot to hand. If urine is passed, make a clean catch (p 188). Inspect the nappy's contents. Examine the genitalia/anus. Find the testes. Rectal examination is rarely needed.
- 11 Note large inguinal nodes. Feel femoral pulses.

Neurological examination After completing the above, much will have been learned about the nervous system; if in doubt, check:

Tone: Passively flex and extend the limbs (provided this will not hurt).

Power and co-ordination: Watch him walk, run, and pick up a small toy and play with it, with each hand in turn. *Reflexes:* Look for symmetry.

Sensation: Light touch and pain testing are rarely rewarding. Fundi.

Ears/throat Leave to the end, as there may be a struggle. Mother holds the child on her lap, one hand on the forehead, holding his head against her chest, and the other round his waist, encircling his arms. Introduce a spatula and get one good look at mouth and tonsils. Inspect the teeth. Examine ear drums (p 528), using the same hold, but held laterally.

Growth Chart height, weight, and head circumference (p 294).

TPR charts Pulse and respiratory rate; T° (rectal T° : normally $<37.8^{\circ}\text{C}$).

Finally ask child and mother if there is anything else you should look at.

Is this child seriously ill? APLS

Recognizing that a child is seriously ill and in need of prompt help is a central skill of paediatrics. It can be uncanny to watch the moment of transformation that this recognition brings to a normally reflective, easy-going doctor who is now galvanized into an efficient, relentless device for delivering urgent care—'omitting nothing necessary and adding nothing superfluous': a frame of mind recommended by Graham Greene for murder, but equally well suited to curing disease. If you are new to paediatrics, take every opportunity to observe such events, and, later, closely question the doctor about what made him act in the way he did, using this page to prepare your mind to receive and remember his or her answers.

- Serious signs** • Consciousness ↓ (not engaging you; apathy; agitation; coma).
 • Hyperventilation or using alae nasae/sternomastoids; wheeze; stridor; apnoea.
 • Ashen, cyanosed, or mottled—or signs of dehydration. medline:9880280
 • Grunting ± chest retractions or head-nodding in time with respirations.
 • Weak/absent pulses, poor capillary refill. To elicit this, press on a digit for 5sec: capillary refill normally takes <2sec. *Don't rely on BP and pulse rate.*
 • High core T°, but cold peripheries.
 • Unreactive or unequal pupils; or decorticate (flexed arms, extended legs); or decerebrate (arms + legs extended); or rigid posture.
 • Weak/high-pitched cry and no spontaneous movement during examination.

Age	Reference interval for:	Breathing rate	Pulse	Systolic BP mmHg
<1yr		30–40/min	110–160/min	70–90
2–5yr		20–30/min	95–140/min	80–100
5–12yr		15–20/min	80–120/min	90–110
>12yr		12–16/min	60–100/min	100–120

Rapid assessment of the seriously ill child (Should take <1 minute.)

- Airway:** • Patency.
Breathing: • Work of breathing *Look:* Rate; recession; accessory muscle use; cyanosis *Listen:* Stridor; grunting; wheeze.
Circulation: • Pulse rate and volume; capillary refill; skin colour and T°
Level of response: • **A=Alert** • **V=**responds to **Voice** • **P=**responds to **Pain**;
 • **U=Unresponsive** (ie 'AVPU'). Also assess pupils & posture.

Causes Commonly sepsis or poisons—also:

- Gastrointestinal: D&V; obstruction (volvulus) • Arrhythmias
- Hypoglycaemia; u&e ↑; metabolic errors • Myocarditis
- Congenital heart disease • Cardiomyopathies
- Intussusception (p 198) • DIC (p 244)
- Haemolytic uraemic syndrome • Reye's syndrome (p 756)

Be alert to rare syndromes, eg toxic shock syndrome (OHCM p 592) or haemorrhagic shock encephalopathy syndrome (HSES; abrupt shock, with bloody diarrhoea, coma, convulsions, hepatomegaly, oliguria, and DIC).

► Action—if very ill • 100% O₂ by tight-fitting mask with reservoir.

- Colloid: 20mL/kg bolus iv; repeat if no better. ≥60mL/kg may be needed.
- Do blood glucose (lab and ward test); u&e (ask specifically for HCO₃⁻, and, if vomiting, Cl⁻); F&C; thick film if tropical travel.
- Crossmatch blood if trauma is possible, or patient looks anaemic.
- Consider the need for CXR, MSU, and lumbar puncture.
- Do swabs, blood culture (+suprapubic urine aspirate if <1yr; before starting blind treatment with iv antibiotics, eg ceftriaxone or cefotaxime, p 258).
- If worsening, insert CVP. Get expert help; do blood gases ± clotting screen.
- Ventilate mechanically, eg if bicarbonate is to be given (pH <7.15).
- If perfusion is still poor (despite 60mL/kg colloid), or CVP >10cmH₂O, dopamine ± dobutamine may be needed. Vasodilatation with nitroprusside (p 281) may rarely be needed to ↑ cardiac output (if the cause is cardiac).

Learning to breast feed

It is better to be apprenticed antenatally to a breast-feeding mother than to rely on simple encouragement and leaflets. www.bmj.com/cgi/content/full/318/7175/30

Reflexes Rooting (searching, with wide-open mouth)→suckling (jaw goes up and down while the tongue compresses the areola against the palate)→swallowing reflex (as milk hits the oropharynx, the soft palate rises and shuts off the nasopharynx; the larynx rises, and the epiglottis falls, closing the trachea).

Skill Don't assume this comes naturally; commonly, learning to breast feed is as hard as learning to drive—and as anxiety-provoking. The best way to learn is from an experienced person in comfortable surroundings—eg sitting in an upright chair, rather than inadequately propped-up in bed. Reassure that a few problematic feeds do not mean that the baby will starve, and that bottle feeding is needed. *Most term babies have plenty of fuel reserves* (earthquake infant-victims may survive for >1 week)—and perseverance will almost always be rewarded. Furthermore, 'top-up' bottle feeds may undermine confidence, and, by altering the GI milieu, diminish the benefits of breastfeeding.

A good time to start breast feeding is just after birth (good bonding; PPH risk↓), but labour procedures may make this hard, eg intrapartum pethidine ± instrumental delivery, T° and BP measurements, washing, weighing, going to a postnatal ward. ► *It is never too late to put to the breast, provided lactation has been maintained.*

Beware of intervening too quickly without observing the mother's efforts. Rather than saying 'that's completely wrong: do it like this. . .' it may be better to say 'good: you and your baby are going to get on fine. One extra tip. . .'

From the baby's viewpoint, breast feeding entails taking a large mouthful of breast-with-nipple, which he or she gets to work on with tongue and jaw. Ensure the baby is close to the mother with the shoulders as well as the head facing the breast—which, if large, may need supporting (mother's fingers placed flat on the chest wall at the base of the breast: avoid the 'scissors' grip which stops the baby from drawing the lactiferous sinuses into his mouth).

- Avoid forcing the nipple into the mouth, eg by placing a hand over his occiput and pressing forwards. Cradle the head in the crook of the arm.
- Explain the signs of correct attachment:
 - Mouth wide open, and chin touching the breast (nose hardly touching).
 - The baby should be seen to be drawing in breast, not just nipple.
 - Lower lip curled back, maximally gobbling the areola (so angle between lips >100°). (Don't worry about how much areola can be seen above the top lip: this gives little indication of where the tongue and lower jaw are.)
 - Slow, rhythmic, and deep jaw movements, as well as sucking movements. (NB: the first few sucks may be fast, shallow, and non-nutritive: here the baby is inducing the mother's 'let-down' reflex, which promotes flow.)
- When helping with placing, it is quite appropriate to 'tease' the baby by brushing his lip over the nipple, and then away. This may induce a nice big gape. With one movement bring to the breast, aiming his tongue and lower jaw as far as possible from the base of the nipple—so his tongue can scoop in the nipple and a good mouthful of breast.
- Keeping on the postnatal ward for a few days, and having the mother learn with an experienced, friendly midwife is also very helpful,⁶ but this facility is now rare in the UK, as cost and other pressures make admissions shorter.

How to express breast milk

It is desirable for every breast feeding mother to learn this skill (access to teaching is *required* before the accolade of 'baby-friendly' can be granted to UK hospitals). There are at least 4 times when expressing is valuable:

- To relieve painful breast engorgement between feeds.
- To keep milk production going when it is necessary to give nipples a rest owing to soreness—which is quite a common problem.
- To aid nutrition if sucking is reduced for any reason (eg prematurity or cleft lip).
- If the mother is going to be separated from her baby for a few feeds, eg going out to work.

The best way to learn is from a midwife, and by watching a mother who is already successfully expressing milk. Pumps may be available. If not, wash hands, and dry on a clean towel, then, try to start flow by:

- 1 Briefly rolling the nipple: this may induce a let-down reflex, especially if the baby is nearby.
- 2 Stroke the breast gently towards the nipple.
- 3 With circular movements, massage the breast gently with the 3 middle fingers.

Applying warm flannels, or expressing in the bath may aid flow, eg while the mother is learning, and only a few drops are being expressed.

Teach the mother to find the 15 or so ampullae beneath the areola: they feel knotty once the milk comes in. Now with the thumb above the areola and the index finger below, and whole hand pressing the breast back on the chest wall, exert gentle pressure on the ampullae. With rhythmic pressure and release, milk should flow (have a sterile container to hand).

Take care that the fingers do not slip down on to the nipple, and damage the narrowing ducts. Fingers tire easily: practice is the key. Concentration is also needed to be sure to catch oddly angled jets.

If kept in a fridge, the milk lasts 24–48h. Frozen milk should be used within 3 months. It is thawed by standing it in a jug of warm water. Any unused milk should be discarded after 24 hours, not refrozen.

Breast feeding and social/biological influences

- Factors which make starting breast feeding harder** • Family pressures, including partner's hostility (10% breast feed vs ~70% if he approves).
• If mother and baby are separated at night in hospital.
• Urbanization, or whenever the mother is expected to work, and there is no convenient place for breast feeding.
• Cultural reframing of breasts as sex objects.
• Rôle-models: if mothers, grandmothers, and elder sisters are not breast feeding, younger sisters may feel embarrassed about starting.

Advantages of the breast

- Breastfeeding gives lower infant mortality rate (wherever there is poverty).
- Colostrum protects from diarrhoeal illnesses. www.txt.co.uk/full.cfm?1024
- Sucking promotes uterine contractions, so avoiding some PPHs.
- Bonding: mutual gaze stimulates emotional input from the mother.
- Breastfed babies have an attractive, sweet smell.
- Breast feeding is clean and cheap and stimulates all five senses.
- Colostrum has endorphins: good for birth-associated stress? medline:11568517
- Breast feeding is cheap, and breast milk is clean.
- IgA, macrophages, lymphocytes (with interferon) and lysozyme protect from infection. Acids in breast milk promote growth of friendly lactobacillus in the baby's bowel. Gastro-enteritis may be less severe if the mother makes and transfers antibodies (an 'immune dialogue').
- GP attendances, otitis media, and pneumonia are rarer if breastfed.
- Breast milk contains less Na^+ , K^+ , and Cl^- than other milk, so aiding homeostasis. If dehydration occurs, risk of fatal hypernatraemia is low.
- Breast feeding reduces the risk of juvenile-onset diabetes mellitus, rheumatoid arthritis, inflammatory bowel disease, and food allergies.
- Breast feeding protects against early-onset atopic eczema and other allergies.
- Breast feeding helps mothers lose weight, and is contraceptive (unreliably!).
- Breast feeding may raise IQ scores (possibly due to long chain fatty acids needed for neurodevelopment). meta-analysis medline~10500022
- Some protection in premenopausal years against maternal breast cancer.

Why is feeding on demand to be encouraged?

- It keeps the baby happy, and enhances milk production.
- Fewer breast problems (engorgement, abscesses).

NB: sleep is less disturbed if baby is kept in bed with mother provided there are no nearby solid objects (eg a wall) against which a baby might be crushed (this may contravene some advice leaflets about SIDS, p 286).

Contraindications to breast feeding • An HIV +ve mother in developed countries • Amiodarone • Antimetabolites • Antithyroid drugs • Opiates.

Problems These include breast engorgement and breast abscess. Treat by using the breast more effectively (better latching-on); aim to keep breasts empty, eg by hourly feeds or milk expression (p 177).

If a breast abscess forms, discard the milk if it is pus-like. Give the mother flucloxacillin 250mg/6h po (it is safe for her baby).

Treat sore nipples by ensuring optimal attachment (p 176), and moist wound healing (eg Jelonet® dressing) not by resting (except in emergencies).

Preterm breast milk is *probably* the best food for preterm infants. Give unheated, via a tube (p 246). Add vitamins D 1000u/day and K (p 246). Phosphate supplements may be needed. Note: even term babies may (rarely) develop rickets ± hypocalcaemia (eg with fits, recurrent 'colds' lethargy, or stridor), if exclusively breast fed, unless vitamin supplements are used (p 208).

Bottle feeding

There are few contraindications to breast feeding but many pressures not to (p 176–8). In many communities over half of mothers are breast feeding at 2 weeks but this reduces to $\leq 40\%$ at 6 weeks. Most change to bottle because of lack of knowledge or no encouragement. Advertising may also have a rôle. The WHO/UNICEF *International Code of Marketing of Breastmilk Substitutes* bans promotion of bottle feeding and sets out requirements for labelling and information on infant feeding. www.babymilkaction.org/regs/thecode.html

Advantages of bottle feeding Fathers can bottle feed. Some mothers value the reassurance of knowing how much milk the baby is taking.

Standard infant formulas (Cow's milk which has been 'humanized' by reducing the solute load and modifying fat, protein, and vitamin content.) As with breast milk, the protein component is whey-based. Several brands are available, eg SMA Gold Cap®. Brands are similar so shopping around for a brand which 'suits' the baby is unlikely to be an answer to feeding problems.

Follow-on formula milks are like standard formulas, but the protein component is casein-based, which delays stomach emptying and allows less frequent feeding. These are marketed to satisfy hungrier babies before they start weaning. Typical age of use: 6–24 months. SMA White® is an example.

Soya milks These contain corn syrup or sucrose, but no lactose, and are indicated in secondary lactose intolerance, galactosaemia, and primary hypolactasia—as well as in vegan families, as they ban all cow's milk. Wysoy® is not the solution for infants with eczema: see p 589. Because of their high aluminium content, avoid soya if preterm, and in renal disease. Soya milk is not indicated in re-establishing feeding (regrading) after gastro-enteritis.¹

Hydrolyzed formula is a cow's milk formula where protein is hydrolyzed to short peptides (eg Nutramigen®). Indications: cow's milk allergy (seen in 1% of babies, eg with bloody diarrhoea and perioral rash) or soya allergy; prevention of atopy (eg in babies with a strong family history). Cow's milk can be reintroduced eg at 1 year (can be risky: so do so in hospital).

Specialist milks Many types exist (eg for gastro-oesophageal reflux, malabsorption, metabolic diseases, etc. Seek advice from a paediatric dietitian.

Preparing feeds Hands must be clean, equipment sterilized, and boiled water used—infected gastro-enteritis causes many deaths in poor countries and considerable morbidity in UK. Powder must be accurately measured. Understrength feeds lead to poor growth and overstrength feeds have, in the past, caused dangerous hypernatraemia, constipation, and obesity.

Feeding Babies require $\sim 150\text{mL/kg/24h}$ ($30\text{mL}=1\text{oz}$) divided over 4–6 feeds depending on age and temperament. Small-for-dates babies may need up to 200mL/kg/day , and large-for-dates babies $<100\text{mL/kg}$. Feeds are often warmed, but there is no evidence that cold milk is harmful. Flow should almost form a stream; check before each feed as teats silt up. The hole can be enlarged with a hot needle. Bottles are best angled so that air is not sucked in with milk.

Weaning 'Solids' should be introduced eg at 4–6 months by offering cereal or puréed food on a spoon. Don't add cereals to bottles. After ~ 6 months follow-on formula may be tried; lumpy food is started so that the baby can learn to chew. Normal supermarket cow's milk may be coped with ~ 1 year old but it may still be too rich in protein, Na^+ , K^+ , phosphorus—and is poor in iron, trace elements, linoleic and alpha-linolenic acids, and vitamins C & B complex. Infants must be able to cope with its higher solute load.¹

Preterm and light-for-dates babies

A neonate whose calculated gestational age from the last menstrual period is <37 completed weeks is preterm* (ie premature).

Low-birth-weight babies fall into 3 groups:

- 1 <2500g but appropriate for gestational age.
- 2 <2500g term infants who are small for gestational age (SGA, ie weight below the 10th centile for gestation—see table).
- 3 Low-birth-weight preterm babies (weight is small for gestational age).

6% of UK infants are <2500g at birth, and 50% of these are preterm. 10% of pregnancies end in spontaneous preterm delivery, and 70% of all perinatal deaths occur in preterm infants (particularly if also growth-retarded).

Causes of prematurity These are mostly unknown. Smoking tobacco, poverty, and malnutrition play a part. Others: past history of prematurity; genitourinary infection/chorioamnionitis (eg *Ureaplasma*)*; pre-eclampsia; polyhydramnios; closely separated pregnancies; multiple pregnancy; uterine malformation; placenta praevia; abruption; premature rupture of the membranes. Labour may be induced early on purpose or accidentally (p 110).

Estimating the gestational age Use the Dubowitz score (p 298).

Small for gestational age babies *Chief causes:* ►Poverty/poor social support may account for 30% of variance in birth weights. medline:11020102
Constitutional/familial factors are also important.

Other causes: Malformation; twins; placental insufficiency (maternal heart disease, BP↑, smoking, diabetes, sickle-cell disease, pre-eclampsia). medline:10895219

The incidence of live-born, very-low-birth-weight infants <1500g is ~0.6%. Neonatal mortality (within 28 days of birth) is ~25%. If the weight is 500–600g only about 10% survive, at best.

Gestational age (based on LMP and ultrasound) is more important for predicting survival than the birth weight alone.

Management If 32 weeks or less, transfer *in utero* to a special centre, if possible. Once born, ensure airway/breathing is optimal; protect from the cold. Measure blood glucose. Take to NICU/SCBU if glucose <2mmol/L. Plan supplemental breast milk or low-birth-weight formula if <2kg. Measure blood glucose before each 3-hourly feed. Tube feed if oral feeds are not tolerated. If oral feeding is contraindicated (eg respiratory distress syndrome) IV feeding is needed (p 246).

Survival if very premature The 'EPICure' study survival figures:

23 weeks' gestation: 12% of live births survived

24 weeks' gestation: 26% of live births survived

25 weeks' gestation: 45% of live births survived.

In other studies, 19% survived at 23 weeks, 40% at 24 weeks, and 54% at 25 weeks. medline-9577278 & 11015506

Mortality is associated with intracranial abnormalities seen ultrasonically.

Disability As a percentage of live births: if 23 weeks' gestation: 5% had no subsequent disability (24 weeks = 12%; 25 weeks = 23%). Morbidity relates to cerebral palsy, squint, and retinopathy (p232). n=150 medline 9577278

Disability may be subtle but specific: one pattern is reduced numeracy if gestation is <30 weeks, from reduced grey matter in the left parietal lobe.¹

*Premature is now used as a synonym for preterm; beware older books, as premature once meant birth weight <5 lbs (2.5kg), before the difference between light-for-dates and preterm was recognized

**Also causes neonatal sepsis, respiratory distress & intraventricular haemorrhage (E&M 1999 4 29)

1 E Isaacs 2001 *Brain* 124 1701–7

See also M Greene 2002 *NEJM* <http://content.nejm.org/cgi/content/short/346/3/146?query=TOC>

Is this baby small for gestational age?

Use centile charts which take into account that first-borns are lighter than subsequent births: the table below gives sample data.

Weeks' station	Tenth centile weight (grams)					
	First born:	boy	girl	Subsequent births:	boy	girl
.....		1220	1260	1470	1340
.....		1540	1540	1750	1620
.....		1830	1790	2000	1880
.....		2080	2020	2230	2100
.....		2310	2210	2430	2310
.....		2500	2380	2600	2480
.....		2660	2530	2740	2620
.....		2780	2640	2860	2730
.....		2870	2730	2950	2810

Common symptoms in infancy

Crying Crying peaks at 8 weeks old (~3h/day, esp. in the evenings). The cries of hunger and thirst are indistinguishable. Feeding at adult-dictated intervals may cause crying in many babies, so demand feeding is preferred. Any thwarting of a baby's wishes may lead to crying, with bouts of screaming. No doubt there are good survival reasons for Nature making babies cry, but in the 20th century this provision may be counterproductive: prolonged crying (as well as lack of parental support) can even provoke child abuse in vulnerable families. Crying may be the last straw for a parent with few reserves. Aim to offer help *before* this stage (eg CRY-SIS self-help group, UK tel. 020 7404 5011). The key skills are not to make parents feel inadequate, and to foster a spirit of practical optimism with one or other parent sleeping and gaining rest whenever the child sleeps.

3-month colic Paroxysmal crying with pulling up of the legs, for >3h on ≥3 days/wk comprise a defining rule of 3s. Cow's milk protein allergy and parental discord and disappointment with pregnancy are implicated—but it may be that such stressed parents may report more colic. Blunted rhythm in cortisol production is reported, suggesting delay in establishing circadian activity of the hypothalamic-pituitary-adrenocortical axis and associated sleep-wake activity. medline:11016553 **Treatment** Nothing helps for long. Try movement (eg carry-cot on wheels, or go for a drive). Trials favour dimeticone (simethicone) drops (40mg/mL 0.5mL po before feeds; avoid if <1 month). Few other drugs are licensed for use at this age. Rarely, chloral is used. In breastfed babies, a cow's milk-free diet for mother is unlikely to help, and a change to soya milk (p 179) in bottle fed babies is often tried, pointlessly, in desperation. Reassure strongly, consider encouraging grandparent involvement. Remember: a crying baby may be a sign of major family relationship problems.

Vomiting Effortless regurgitation of milk is very common during feeds ('possetting'). Vomiting between feeds is also common. >1 forceful vomit per day usually indicates some pathology, eg gastro-enteritis; pyloric stenosis; hiatus hernia/reflux (p 272 mucus ± blood in vomit, which is usually effortless); pharyngeal pouch; or duodenal obstruction (there is bile in the vomit). Observing feeding is helpful in deciding if the vomiting is projectile (eg over the end of the cot), suggesting pyloric stenosis.

Nappy rash or diaper dermatitis 4 types:

- 1 The common ammonia dermatitis—red, desquamating rash, sparing skin folds is due to moisture retention, not ammonia. It often responds to frequent changes of nappy, or nappy-free periods, careful drying, and an emollient cream. Avoid tight-fitting rubber pants. 'One way' nappies may be helpful at night (eg Snugglers®). medline:1101357
- 2 Candida/thrush is isolatable from ~½ of all nappy rashes. Its hallmark is satellite spots beyond the main rash. Mycology: see p 590. Treatment as above, + clotrimazole (± 1% hydrocortisone cream, eg as Canesten HC®). One trial favours thrice-daily mupirocin ointment. N 20 medline:1048745
- 3 Seborrhoeic eczematous dermatitis: a diffuse, red, shiny rash extending into skin folds, often associated with other signs of a seborrhoeic skin eg on occiput (cradle cap). Treatment: as for 1. medline:621188
- 4 Isolated, psoriasis-like scaly plaques (p 586), which can be hard to treat.

Agents to avoid: Boric acid and fluorinated steroids (systemic absorption); oral antifungals (hepatotoxic); gentian violet (staining is unpopular)

Sleep problems See p 390.

Chronic childhood illness, and family support

Diseases such as severe asthma, CNS disease, and neoplasia may cause disintegration of even the most apparently secure families: ► *consequent strife and marital breakdown may be more severe and have more far-reaching consequences than the illness itself.*

Remember that illness makes families poor, and movement down the social scale may lead to unpredictable consequences in the fields of housing and (un)employment.

Marital disharmony may seem to be beyond the scope of paediatrics, but any holistic view of child health *must* put the family at the centre of *all* attempts to foster child health and wellbeing.

We see many families coping admirably with severe, prolonged illness in a child. But do not presume that because things are ok in clinic today, you can afford to neglect the fostering of family life. Given a certain amount of stress almost *all* families will show psychopathology, in time. Your job is to delay this. Counselling skills are frequently needed (p 376)—but do they work? Various tools are helpful in answering this question:

medline:8916632

- Communication is vital in any family, and this can be measured by scales such as the Communication Skills Test (CST).*
- The Dyadic Adjustment Scale (DAS) looks at the emotional interaction within a marriage or any other pairing.
- The Miller Social Intimacy Scale (MSIS) looks at intimacy in relationships, and intimacy is, beyond doubt, a source of strength within a family experiencing illness in a child.

medline:1640317

<http://newton.uor.edu/FacultyFolder/Rickabaugh/Methods/Measures.html#Relationships>

- Specific therapies validated in randomized trials in families coping with severe chronic illness in a child include Emotionally Focused Marital Therapy (EMT), with these benefits:

medline:9373753

- Higher overall levels of marital adjustment (on the DAS scale).
None of the couples receiving the active therapy deteriorated by >7 points, whereas one-third of couples in the control group had such a deterioration (which shows the stress these families were under, and supports the notion that family psychopathology in the face of chronic childhood illness is an evolving phenomenon—the stress is reversible if you take certain steps).
- Better levels of intimacy as measured on MSIS scale, with effects persisting for at least 5 months.
- Lower rates of negative communication (on the CST score).
- NNT = 2 (2 couples need treating for one to improve).

E-BM 2 46

*Full Instruments available in: K Corcoran & J Fischer J (1987). Measures for clinical practice: A sourcebook. New York

Growth, failure to thrive, and short stature

Is growth normal? is a key question in determining the health of a child. Take any opportunity to weigh and measure a child. A series of plots on centile charts (p 296) shows if growth is slow (growth curve crosses the centiles). NB: the growth rate in mid-childhood is 5–6cm/yr; this accelerates at puberty (peak height velocity), before epiphyses start to fuse.

Failure to thrive means poor weight gain in infancy (falling across centile lines). Head circumference is preserved relative to linear growth, which is preserved relative to weight. In 95% this is due to not enough food being offered, or being taken. In developing countries, poverty is the main cause; in the UK it is difficulty at home, emotional deprivation, or unskilled feeding. Other causes: see below. NB: the cause may simply be idiosyncratic growth pattern; normal child of short stature (prematurity, ↓ birth weight, short siblings or parents)—particularly if he is happy and contented.

In the consultation:

- Signs of abuse (p 284)
- Feeding patterns
- Behaviour; activity level
- Family finances
- Dysmorphic face
- General health & happiness
- Chart family heights
- Parental illness?

Be sceptical about reliability of data. Was the child clothed during weighings? Length measurements are particularly prone to error: growth velocity is more useful than measurements taken at a single time.

Issues to address: • Feeding and maternal interaction. Is the child anorectic or ravenous—‘hyperphagic short stature’? medline:8709732

- If breast feeding, does he get a good mouthful of breast? (p 176)
- If bottle feeding, does the teat’s hole allow milk to flow through?
- Does weight gain return if the child is removed from the family?
- Is there evidence relevant to child protection proceedings?

Tests: It is a great skill to know when to investigate. It’s better from the child’s point of view to ask a trusted colleague’s advice before painful tests. In one study only 39 of 4880 tests were helpful. medline:8298353

MSU (expect false +ves if bags are used; but avoid routine suprapubic aspirations); U&E/glucose, LFT, Ca^{2+} , proteins, immunoglobulins, CRP, TSH; FBC; sweat test; urinary amino ± organic acid chromatography; stools (MC&S ± sugar detection); CXR, renal or CNS ultrasound, skeletal survey for dwarfism and abuse; jejunal biopsy; ECG/echo. In *non-organic failure to thrive*, studies favour weekly visits from trained lay visitors. medline:7539121

Short stature is a height below the 3rd centile (p 296). If the parents are both small, their child will be likely to be small too (constitutional short stature accounts for ~80% of short children). Use the method shown on the charts to correct for mid-parental height. ► *Any chronic disease can cause short stature.* Hypopituitarism (an important cause of short stature) usually manifests after age 2yrs: look for relative obesity, without any other explanation for low growth velocity (ie < 25th centile; measure for ≥ 1yr, see p 296). Deficiency of growth hormone (GH) is shown by an impaired rise (peak GH < 15mu/L) after a stimulus (eg sleep or hypoglycaemia, induced by iv insulin, *OHCM* p 308). Pre-school screening for short stature is the aim. To be effective synthetic GH should be given early. Growth hormone dose example: 0.5–0.7u/kg/week sc, perhaps more during puberty. Daily doses are probably better than thrice weekly regimens. Other pituitary hormones may also be deficient (*OHCM* p 308).

Other causes of short stature

Poverty:	Endocrine:	Drugs:	Genetic:
Poor or ineffective diet (also coeliac)	Hypothyroid	Steroids	Turner’s
Physical or psychological abuse	GH ↓ (see above)		Cystic fibrosis
			Noonan sy, p 754

Causes of height ↑: Thyrotoxic; precocious puberty; Marfan’s; homocystinuria.

ethnospecific growth charts

medline 8622378

It is clear that some populations are inherently shorter than others, and this poses problems when using growth charts. Consider these facts: The Dutch are the tallest *nation* on earth (mean ♂ height = 1.84m): the smallest *population group* is the Masai people (eg in Tanzania and Kenya). African and Afro-Caribbean 5–11yr-olds height is ~0.6 standard deviations scores (SDs) greater than white children living in England. Gujarati children and those from the Indian subcontinent (except those from Urdu- or Punjabi-speaking homes) have heights ~0.5 SDs less than white children living in England. Gujarati children's weight-for-height is ~0.9 SDs less than expected for Afro-Caribbeans, or white children in England—so Gujarati children's weight is ~1.5 SDs less than for white children living in England. Urdu and Punjabi weight is ~0.5 SDs < expected for white UK children. Published charts have centile lines 0.67 SDs apart; for height and weight shift the centile lines up by 1 centile line division for Afro-Caribbeans. Re-label Gujarati children's weight charts, so the 0.4th centile becomes the ~15th centile, and the 2nd weight centile becomes the ~30th centile. For most other Indian subcontinent groups, height & weight need shifting down, eg relabel the 0.4th & 2nd centile lines 1.5th & 6th respectively. NB: Sikh children are taller and heavier than caucasians. medline:7979492 Body-mass index centiles are said to be appropriate for Afro-Caribbeans, but recalculate as above for Indian subcontinent children, except for Gujarati speaking children (0.4th & 2nd centiles → 4th & 14th). Trends towards tallness with each generation occur at varying rates in groups, so 3rd generation immigrants are taller than expected using 2nd generation data. Intermarriage adds further uncertainty. NB: to print the new CDC charts, see www.cdc.gov/nchs/about/major/nhanes/growthcharts/clinical_charts.htm The 1990 standard UK charts have recently been validated. medline:10999860 NB: ↑weight of the school children is a big public health issue.

Celiac disease

Malabsorption typically presents with diarrhoea, failure to thrive ± anaemia (folate↓; ferritin↓), possibly with abdominal protrusion, everted umbilicus and wasted buttocks (if late-presenting). As subclinical/latent forms exist, investigate any unexplained anaemia, fatigue, 'irritable bowel' symptoms, diarrhoea, weight↓, arthralgia, eczema, and short stature. Patients may present at any age.

Cause: Enteropathy induced by gluten (in wheat, barley, rye, and oats).

Diagnosis: Screen with IgA gliadin, anti-reticulín, & anti-endomysium antibodies; specificity ≈100%, if there is no IgA deficiency (coeliacs are at risk of this, so measure this too, + CRP & ferritin medline:11782119). Confirm by finding villous atrophy on small bowel biopsy (endoscopic under GA is better than Crosby capsule). Villi return to normal on the special diet; avoid a gluten challenge test, unless diagnosis is in doubt. medline~9888912

Treatment: *Gluten-free diet:* no wheat, barley, rye, or oats (hence no bread, cake, or pies). Rice, maize, soya, potato, and jam are OK. Gluten-free biscuits, flour, bread, and pasta are prescribable. Minor dietary lapses may matter. After 5yrs: cautious reintroduction of gluten may be tried.

Other causes of malabsorption: Cystic fibrosis; post-enteritis enteropathy; giardia; rotaviruses; bacterial overgrowth; milk sensitivity; worms; short bowel syndrome. *Coeliac associations:* Diabetes mellitus (type I).

Diarrhoea

Deaths/yr = 3×10^6 children, worldwide

Faeces are sometimes so liquid they are mistaken for urine. NB: it is normal for breast fed babies to have liquid stools. Some cow's milk causes harmless green stools. Diarrhoea may be an early sign of any septic illness.

Gastroenteritis The big danger from small intestine infection is dehydration ± U&E imbalance (p 250) from D&V. Rotavirus causes 30% of deaths. medline:11190969 There is often associated otitis media ± coryza. Other enteric viruses: astrovirus, small round-structured viruses (SRSV, eg Norwalk agent), calicivirus. **Treatment:** ► If dehydrated, see p 250. Weigh the child (to monitor progress) and to quantify the level of dehydration, if a recent previous weight is available. Stop bottle milk (but if breast feeding, continue) and solids, and give oral rehydration mixture, eg Rehidrat® or Dioralyte®, which is 60mmol Na⁺/L with an osmolarity of 240mmol/L, and is better in non-cholera diarrhoea than standard WHO solutions (90mmol & 311mmol/L respectively). N=447 medline:7837862 & 9002327 & 8551428

Milk is re-introduced after 24h (avoid regrading, even if <6 months), or sooner if the child recovers and is hungry: starvation delays recovery. N=159 medline:8093495

NB: breast feeding supplies antibodies and maintains milk production, but in poor communities breast milk is likely to be Zn-deficient, and Zn supplements + vit. A help stop diarrhoea becoming chronic (eg 20mg Zn/24h po for 14d). N=800 medline:11498488

Complications: Dehydration; malnutrition; temporary sugar intolerance after D&V with explosive watery acid stools. (Rare; manage with a lactose-free diet.) Post-enteritis enteropathy resolves spontaneously after 6–8 weeks.

Tests: Stools (look for bacteria, ova, cysts, parasites). **Prevention:** Hygiene, good water and food, education, fly control. Rotavirus vaccines are being assessed.

Secretory diarrhoea Causes: Bacteria (campylobacter, staphylococcus, *E. coli*) and, where sanitation is poor, salmonella, shigella, and *Vibrio cholerae*; giardiasis; rotavirus; amoebiasis; cryptosporidium. Inflammatory bowel disease.

Toddler diarrhoea 'Like peas & carrots'; due to intestinal hurry; resolves by 4yrs

Other causes Food intolerance (coeliac disease; cows' milk or wheat, galactose, glucose, sorbitol, or lactose intolerance); to find reducing substances: mix 5 drops of stool with 10 drops H₂O + 1 Clinitest® tab); antibiotics; deficiencies (zinc, copper, Mg²⁺, vitamins); kwashiorkor. medline:8892186

Causes of bloody diarrhoea Campylobacter; necrotizing enterocolitis (in neonates); intussusception (often <4 years old); pseudomembranous colitis; haemolytic uraemic syndrome (p 280); inflammatory bowel disease (rare, even in older children: look for weight↓; anaemia; WBC↑; platelets↑; ESR/CRP↑).

Malnutrition

WHO site: www.txt.co.uk/sum.cfm?103

As a major cause of mortality/morbidity worldwide, this has a global importance to us all. **Kwashiorkor** is due to ↓ intake of protein & essential aminoacids. It causes growth retardation; diarrhoea; apathy; anorexia; oedema; skin/hair depigmentation; abdominal distension; glucose↓; K⁺↓; Mg²⁺↓; Hb↓; cholesterol↓; albumin↓. **Treatment** depends on re-education (of child, family, politicians) as well as on offering a gradually increasing, high-protein diet with vitamins.

Marasmus is deficiency of calories, with ↑ discrepancy between height and weight. It is HIV-associated. medline 10824214 **Presentation:** Abdominal distension; diarrhoea, constipation, infection. **Tests:** Serum albumin↓. In young children a mid-arm circumference <9.9cm (any age) predicts severe malnutrition better than being <60% of median weight for age, or 85% of median height for age, and 70% of median weight for height. **Treatment:** Parenteral feeding may be needed to restore hydration and renal function. Next offer a balanced diet with vitamins. Despite this, stature and head circumference may remain poor. Kwashiorkor and marasmus may occur together (protein-energy malnutrition).

Southern diarrhoea is only an excuse for Northern amnesia

As you read this page two events unfold: in the Northern hemisphere a child is born with a silver spoon in his mouth—his future assured thanks to incubators, ventilators, wealth, and family planning; and, diametrically opposed to this birth another occurs in the Southern hemisphere where the silver was mined for that silver spoon. This child, according to our stereotype must 'Wait for his future like a horse that's gone lame—to lie in the gutter and die with no name.' We assume that this death is from diarrhoeal diseases that we are all working, more or less efficiently, towards controlling—and we are pleased to blame non-human agencies for these deaths. This model of our imperfect world does not stand up to scrutiny for two reasons: one is easy to understand (the diarrhoea was the mode of dying not the cause of death, which was poverty) and the other is impossible to understand—we didn't just let him die. We wanted him to die: in fact we killed him: in some cases, literally. Follow this well-documented thought sequence: There are too many homeless children living in my back yard → This threatens health and hygiene → How do we deal with this? → Other threats to health and hygiene are vermin → Street children are a sort of vermin → Vermin need eradicating → Let's shoot the vermin → Extrajudicial shooting of children—as occurred in July 1993 in Rio de Janeiro. Most 'ordinary decent folk' approved of the killings when they phoned a local radio station, as the events unfolded. . . 'I killed you because you had no future.'

medline:9654281

The point of all this is to illustrate that if we want to do something for children it is no good just doing something about the big killers, such as diarrhoea, and it's no good simply attacking poverty, for there is something dark in our human heart which needs addressing before purely statistical or biological interventions have a chance of success. There is only one way of influencing human nature, and that is through dialogue. So in this sense, the treatment of diarrhoea is dialogue.

Childhood urinary tract infection (UTI)

Presentation ▶ Often the child may be *non-specifically* ill. Infants may present with collapse and septicaemia, and toddlers as 'gastro-enteritis', failure to thrive, colic, or PUO. Many with dysuria and frequency have *no* identifiable UTI, and will often have vulvitis. The urinary tract is normal in most with UTI but ~35% have vesico-ureteric reflux (VUR), ~14% have renal scars (most will have reflux too), ~5% have stones, ~3% will develop hypertension. Each year in the UK, 10–20 children enter endstage renal failure programmes because of complications from reflux and chronic pyelonephritis.

Definitions *Bacteriuria*: Bacteria in urine uncontaminated by urethral flora. It may be *covert* (few, unreported symptoms), and can lead to renal scarring, BP ↑ and, rarely, chronic renal failure. *UTI* denotes symptomatic bacteriuria, that may involve different GU sites (check for loin and suprapubic tenderness; ask about fever and dysuria). *Chronic pyelonephritis* is a histological/radiological diagnosis. Juxtaposition of a cortex scar and a dilated calyx is the key to its diagnosis. It is a big cause of hypertension and can result in renal failure, eg if the kidneys are congenitally dysplastic. During micturition, urine may reflux up the ureters, seen on a micturating cystogram (requires catheterization) or MAG scan (catheterization not needed)—grading: **I** Incomplete filling of upper urinary tract, without dilatation. **II** Complete filling ± slight dilatation. **III** Ballooned calyces. **IV** Megaureter. **V** Megaureter + hydronephrosis.

UTI incidence Boys: ≤0.23%/yr; girls: 0.31–1%. Sex ratios are reversed in neonates. **Recurrence**: 35% if >2yrs old. **Prevalence of covert bacteriuria** in schoolgirls: ~3%. **Prevalence associated GU abnormalities**: 40% (½ have reflux, others: malpositions, duplications, megaureter, hydronephrosis). **Renal scars and age**: We used to concentrate on treating babies early, thinking new scars were rare after 4yrs old, but prospective ^{99m}Tc dimercaptosuccinate (DMSA) scintigraphy (the best test) shows new scars appearing on repeat scans in 43% of those <1yr old, 84% of those aged 1–5, and 80% of those >5yrs old.

Tests *Urine collection*: In infants, *suprapubic aspiration* is definitive (and organisms are significant) but invasive. An alternative is to wash genitals gently with water and tap repeatedly (in cycles of 1min) with 2 fingers just above the pubis, 1h after a feed, and wait for a *clean catch*, avoiding the first part of the stream. Do prompt microscopy and culture. >10⁸ organisms/L of a pure growth signifies UTI. For suprapubic aspiration, ward ultrasound helps identify a full bladder. **Method**: clean skin over the bladder; insert a 21–23G needle in the midline 2½cm above symphysis pubis. Aspirate as you advance the needle. Other devices: *dipslides* (go out of date rapidly), *culture pads* (Microstix®). *Urine bags* applied to the perineum cause many false +ves, and urine collection on *sanitary towels*, via a syringe, is much cheaper and perhaps preferable.

Ultrasound (us): us is cheap and noninvasive, and worthwhile even in 1st UTI: sensitivity 43%; specificity 99%; but reflux and scarring may be missed in up to 11%. medline.10799427 Most will be in infants, or occur with fever and vomiting, or be with *recurrent UTI*, so if these are present and us scan is normal, proceed not to IVU, which is radiation-rich and unreliable, but to:

⁹⁹Technetium renography—static for scarring (^{99m}Tc DMSA scan), dynamic for obstructive uropathy ± *isotope cystography*. *Micturating cystourethrography (MCU)* is still the best way of excluding reflux. In general, it is *not* needed if initial tests are normal, pyelonephritis is unlikely, there is no family history of reflux, and there are no *recurrent UTIs*; it is invasive and unpleasant, but careful preparation with play therapy (p 384) mitigates this. If it is done ill-advisedly on an uncomprehending and angry toddler, it may constitute assault. NB: operating on reflux is unlikely to improve renal function. J Smellie 2001 Lancet 357 132

Treatment and prevention of urinary infections

Antibiotics: • Trimethoprim 50mg/5mL, 50mg/12h po for 7 days* (halve dose if <6 months old; double if >6yrs), nalidixic acid, or co-amoxiclav
• Avoid constipation • ↑ Oral fluids • Encourage full voiding • Repeat msu.

Treatment of reflux: If prophylactic antibiotics fail, ureteric reimplantation can reduce reflux, but scarring remains. Keep on antibiotic prophylaxis.

Prevention Just one episode of reflux of infected urine may initiate renal scarring, so screening for bacteriuria is useless: damage happens too quickly. But once a UTI is suggested (eg by microscopy) treat it *at once*, whatever the age, before you know culture sensitivities, as renal damage may be about to happen, while at the same time doing all you can to ensure a high-quality urine sample is analysed. Consider trimethoprim prophylaxis (1–2mg/kg/24h at night), while awaiting surgery or investigations—and sometimes indefinitely (the optimum duration is unknown, but may be after 2 –ve cystograms, if the indication is reflux).* Consider screening siblings for reflux.

Surgical correction of moderate reflux is 'unlikely to be beneficial', and in minor reflux is 'likely to be harmful' (carefully constructed EBM phrases!)

*J Larcombe 1999 *BMJ* i 1173 www.txt.co.uk/full.cfm?1048

Acute lymphoblastic leukaemia (ALL)

ALL is the commonest childhood leukaemia; other forms: see *OHCM* p 656–9.

Causes One model invokes genetic predisposition with later exposure to triggers such as radiation, benzene, HTLV-1, or abnormal response to infection.¹

Classification *Common ALL* (75%): Blasts have characteristics of neither T (rosette) nor B lymphocytes, but have a characteristic surface polypeptide. Median age: 4yrs. Prognosis: good (86% 5yr survival).

B-cell ALL. Blasts look like those of Burkitt's lymphoma. Poorer prognosis (but now 70% disease-free survival). Subtypes: •Early pre-B, eg with cytoplasmic monoclonal antibodies (CD22+, CD79a+, CD19+) and surface immunoglobulins (slg); •Pre-B-cell; •Transitional pre-B-cell; •B-cell (slg+, slg_k+, or slg_λ).

T-cell ALL: Median age: 10yrs. Prognosis: ~40–50% disease-free survival.

Null-cell ALL: Blasts show none of the above features.

Classification by ploidy: Hypodiploid (7% of ALL patients), diploid (8%), pseudodiploid (42%), hyperdiploid with 47–50 chromosomes (27%) or hyperdiploid with >50 chromosomes (27%), or triploid/tetraploid (1%).

Worse prognosis if: Black, older, male, wcc >100 × 10⁹/L; Philadelphia translocation [t(9;22)(q34;q11)]—seen in 12% (0–15% disease-free at 5yrs).

Presentation Pancytopenia (pallor, infection, bleeding), fatigue, anorexia, fever, bone pain. The period before diagnosis is often brief (2–4 weeks).

Tests wcc↑, ↓ or ↔. Normochromic, normocytic anaemia ± platelets↓. Marrow: 50–98% of nucleated cells will be blasts. csf: pleocytosis (with blast forms), protein ↑, glucose ↓. Biochemistry: urate↑, LDH↑.

Treatment In the UK, almost all children are treated according to the MRC/UK-ALL protocol. This entails remission induction (± a briefly intensified treatment to eradicate residual blast cells *medline:10720126*), CNS prophylaxis, and maintenance therapy. Risk assessment ensures only those most likely to relapse get the most aggressive 4-drug treatment with preventive meningeal irradiation (eg if presenting wcc >50 × 10⁹/L; age >9yrs; T-cell immunophenotype; pre-B-cell with the t(1;19) translocation). Pneumocystosis is prevented by co-trimoxazole 10mg/kg/12h po. While the wcc is <1.0 × 10⁹/L use a neutropenic regimen. **Drugs**: High-dose cyclophosphamide, cytarabine, methotrexate (intrathecal), mercaptopurine, asparaginase, daunorubicin, etoposide, tioguanine, vincristine, prednisolone. NB: life-threatening myelosuppression occurs if deficient in thiopurine methyltransferase (so consider pre-R_x pharmacogenomic analysis).

Reinforce remission by use of non-cross-resistant drug pairs, to reduce the leukaemic clone, and ↓frequency of drug-resistant mutants and, therefore, relapse. Bone marrow transplants are not routinely used, but may be needed in those at highest risk, eg Philadelphia chromosome, wcc >200 × 10⁹/L, MLL gene rearrangement, B-cell ALL with the t(8;14) translocation. [•] *medline:11001892*

Much treatment, including iv cytotoxics, is now given at home.

Prognosis 75% 5yr event-free survival. **Treatment related deaths**: 2% (UKALL xi). Deaths during induction: 1% (chiefly bacterial and fungal infections). Deaths in remission: 1% (eg measles and *pneumocystis carinii*). *medline:11167821*

Pitfalls •Serious infection with no tell-tale fever or tachycardia—eg: zoster, measles, CMV, candidiasis, pneumocystosis. Sepsis management: p 191 & 248.

- Hyperuricaemia at induction after massive cell destruction. Prevent by pre-treating with a high fluid intake and allopurinol 10–20mg/kg/24h.
- Ignoring the patient's quality of life.
- Omitting to examine the testes (a common site for recurrence).
- Inappropriate transfusion (leukostasis if wcc >100 × 10⁹/L). *medline:10975379*
- Failing to treat side-effects of chemotherapy optimally—eg 5-HT₃ receptor antagonists such as ondansetron are better than other anti-emetics.
- Drug-induced delayed cancer elsewhere (risk of CNS tumours or secondary acute myeloid leukaemia is ~3–4%). Risk assessment allows lower doses.

febrile neutropenic patients who may be septicaemic (eg from an infected venous catheter)

suspect infection when untoward events take place in a neutropenic patient ($wcc < 2 \times 10^9/L$, or neutrophils $< 1 \times 10^9/L$). Temperature should be monitored often. Transitory rises ($\leq 38^\circ C$) may be ignored if lasting for 6h only if the child is well. The importance of parental (and child) education, and of swift routes to hospital need emphasizing.

Do blood cultures and MSU; swab all orifices. Do FBC, CRP & serology. Get expert advice from your senior, and a microbiologist.

Blind treatment will depend on local protocols, and may include imipenem, the broadest spectrum beta-lactam antibiotic, which may have advantages over cephalosporins if an anaerobe such as *Bacteroides* is suspected (NB: imipenem is not indicated in CNS infections). medline:10689719

imipenem—Dose in children $< 40kg$: 15mg/kg/6h max 2g/24h. Heavier children have the adult dose: 12.4mg/kg/6h ivi; max dose, eg 2g/24h. Do U&E. If creatinine clearance $< 70mL/min$, ↓dose frequency (see Data sheet). NB: do not use IM formulations ivi, and the IM formulation is not to be used in children. If blood culture does prove +ve, either change imipenem after microbiological advice, or continue it for 5 days, provided he or she has been afebrile for $> 24h$. If blood cultures are -ve, give for a few days and send home when well and afebrile for $> 24h$.

imipenem SEs: (It is usually well-tolerated.) Thrombophlebitis, anuria, polyuria, seizures (eg in ~2%), confusion, psychic disturbance, encephalopathy, vertigo, tinnitus, transient hearing loss, BP↓, pruritus, taste perversion, pseudomembranous colitis; arthralgia; eosinophilia; $wcc \downarrow$; Hb↓; LFTs↑. There are no clinically important drug interactions. medline:11033734

Meropenem is preferred in some units. It is more active against most Gram -ve pathogens than imipenem but is less active against most Gram +ve ones. medline:10776838 Dose: 3 months–12 years: 10–20mg/kg/8h ivi over 30mins; over 50kg weight: adult dose (eg 1g/8h ivi). SE: D&V (eg antibiotic-associated colitis); abdominal pain; LFTs↑; platelets↓; ↓partial thromboplastin time; +ve Coombs' test; eosinophilia; neutropenia; headache; paraesthesia; rash; pruritus; convulsions (rare). medline:11389117

If fever persists and blood cultures remain -ve ask: Is *aciclovir* indicated? Is a fungus possible (for *amphotericin* see Data Sheet & OHCM p 166).

Teicoplanin For blind treatment of the worst infections, consider adding this to imipenem. Dose: 6–10mg/kg/12h iv for 3 doses, then daily (neonates: 16mg/kg on day 1, then 8mg/kg/day). SE: (few): dizziness; headache; $wcc \downarrow$; platelets ↓/↑; LFTs↑; allergy (rare). Interactions: none. medline:10949982

Cystic fibrosis

This is one of the commonest autosomal recessive diseases (~1/2000; ~1/2 of Caucasians are carriers); it reflects mutations in the cystic fibrosis transmembrane conductance regulator gene (CFTR) on chromosome 7, which codes for a cyclic AMP-regulated chloride channel. There is a broad range of severity of exocrine gland function, leading to meconium ileus in neonates (and its equivalent in children), lung disease akin to bronchiectasis, pancreatic exocrine insufficiency and a raised Na^+ sweat level—depending in part on the type of mutation (often ΔF_{508} ; but other mutations, eg in intron 19 of CFTR, cause lung disease but no increased sweat Na^+).

Antenatal (p 210) carrier-status testing is possible, as is pre-implantation analysis after *in vitro* fertilization: at the 8 cell stage, 1 cell is removed from the embryo, and its DNA analysed; only embryos without the cystic fibrosis gene are offered for implantation—this may be more acceptable than terminating fully formed fetuses after chorionic villous sampling.

Diagnosis 10% present with *meconium ileus* as neonates. Most present later with *recurrent pneumonia* (\pm clubbing), *steatorrhoea* (if $>7\text{g/day}/100\text{g}$ of ingested fat), or *slow growth*. **Sweat test:*** Criteria for +ve test: $\text{Na}^+ >60\text{mmol/L}$ and sweat $\text{Cl}^- >70\text{mmol/L}$ on 2 occasions (inaccurate if $<100\text{mmol}$ of sweat is collected or $\text{Na}^+:\text{Cl}^-$ discrepancy $>20\text{mmol/L}$). medline:240323 The test is difficult, so find an experienced worker (false +ves in dehydration, endocrine diseases, if on certain antibiotics, and in eczema). Sweat is collected on to filter paper fixed to the forearm. See OTM 3e p2750 [2539112]. **Other tests:** Genetic tests;*:** CXR: shadowing suggestive of bronchiectasis, especially in upper lobes; malabsorption screen; glucose tolerance test; lung function tests; sputum culture. Mycobacterial colonization affects up to 20%; consider if rapid deterioration.

Treatment ▶ Offer genetic counselling to every family (p 212). Long survival depends on antibiotics and good nutrition. **Respiratory problems:** Physiotherapy ($\times 3/\text{day}$) must start at diagnosis. Teach parents percussion + postural drainage. Older children learn the forced expiration technique. Organisms are usually *Staph aureus*, *H influenzae* (rarer), and *Streptococcus pneumoniae* in younger children. *Pseudomonas aeruginosa* acquisition often late, but eventually $>90\%$ are chronically infected. *Berkholderia cepacia* (*Ps cepacia*) is associated with rapid progression of lung disease (prompt diagnosis using polymerase chain reaction on DNA samples may be available: isolate patient). Treat acute infection after sputum culture using higher doses, and for longer than normal. If very ill, ticarcillin (50–60 mg/kg/6h iv) + netilmicin (p 238), or ceftazidime (50mg/kg/8h iv) alone may be needed before sputum results are known. Admissions may be reduced by using nebulized ticarcillin and tobramycin at home. If reversible airways obstruction is a problem, give inhaled salbutamol (look for *Aspergillus* in sputum). Lung transplant (heart + lung, or double lung) works for some, with good results limited by donor availability (avoid raising hopes). Ensure pertussis, MMR, and pneumococcal vaccination.

Gastrointestinal problems and nutrition: Energy needs rise by ~130% because of chronic lung inflammation. Most have *steatorrhoea* from pancreatic malabsorption and will need enzymes: Pancrex V® powder mixed with food for infants—and Pancrex V Forte® for older children, ≤ 1 tabs/meal—to give regular, formed, non-greasy bowel actions. Most older children have enzymes in microspheres (eg Pancrease®, Creon®) so fewer tablets are needed. Cimetidine 10mg/kg po $\frac{1}{2}$ h before each meal may also be needed. If this controls *steatorrhoea*, a low-fat diet is not

needed, but vitamin supplements are still required (A & D, eg as Abidec® 6mL/24h po for infants or as multivitamin capsules 2/24h po for older children). Diet should be high calorie/high protein. Fine-bore nasogastric feeding is needed only if weight cannot otherwise be maintained.

If GI obstruction occurs (eg if Pancrex® is omitted), urgent admission to specialist hospital is needed for medical treatment (laparotomy is contraindicated unless perforation is imminent).

Diabetes mellitus/impaired glucose tolerance: Risk increases with age and higher if homozygous for the $\Delta F508$ mutation. Insulin may be needed. Adjust the dose to the (optimized) diet, not vice-versa. Only try oral hypoglycaemics if nutrition is satisfactory. medline:11220706

Psychological help: Parents and children need expert counselling. The Cystic Fibrosis Research Trust and regional centres may help here.

Complications •Pneumonia (resistant organisms) •Haemoptysis
Nasal polyps •Pneumothorax (in 20%) •Diabetes \pm cirrhosis
♂ infertility (stenotic or absent vas deferens) •Cholesterol gallstones
Pulmonary osteoarthropathy •IV therapy problems
Fibrosing colonopathy (thought to be related to enzyme dose).

Meconium ileus Presents with vomiting in the 1st 2 days of life. Distended loops of bowel may be seen through the abdominal wall. A plug of meconium may show as a firm mass in one such loop. In most other causes of GI obstruction, lateral decubitus films will show fluid levels. This is not the case in meconium ileus. Tiny bubbles may be seen in the meconium. **Treatment:** Place a nasogastric tube. Seek expert advice. Washout enemas may be needed, as may surgical excision of the distal ileum (or whatever contains the most meconium). UK cystic fibrosis society:tel. 020 8464 7211

Prognosis Death may be from pneumonia or cor pulmonale. Most survive to adulthood (median survival is now 30yrs). 5-year survivorship models take account of forced expiratory volume in 1sec as a percentage of predicted normal, gender, weight-for-age z score, pancreatic function, plasma glucose, *Staph aureus* & *Burkholderia cepacia* infection, and annual number of acute pulmonary exacerbations. medline:11207152

The future This may lie in agents to reduce viscosity of sputum with synthetic human DNA-ase which fragments viscous DNA from dead neutrophils in sputum (an example, with modest clinical effect, is Dornase alfa: it has been tried in a once-daily dose via an air compressor with a jet nebulizer).

Lung transplantation is getting safer; considered in those who are deteriorating ($FEV_1 < 30\%$ of expected) despite maximum therapy, provided nutrition is good, and there is no active mycobacterial or aspergillus infection.

Gene therapy aims to deliver normal copies of the cystic fibrosis gene into patients, so allowing them to make CFTR protein. Viral vectors and liposomes have been used to get the gene into cells. medline:11216908

In a sense, the future is already here: but for most it's just too expensive.

Normal sweat Cl values don't exclude cystic fibrosis. [7533604]. In those borderline sweat test and a Cl/Na ratio ≥ 1 CF should be considered. But a Cl/Na ratio < 1 doesn't exclude CF, since patients carrying mild mutations may have sweat Na levels higher than those of Cl. [8649916] Infants with sweat Cl levels of 40–60mmol/L require further investigation. [10821727]

Having 2 mutations is associated with severe disease ($\Delta F508$, W1282X, G542X, N1303K, 1717-G \rightarrow A). Carrying one mutation may not be so bad (3849 + 10 kb \rightarrow A T). [8649916]

Rheumatic fever

This is a systemic febrile illness caused by a cross-sensitivity reaction to Group A β -haemolytic streptococcus, which, in the 2% of the population that is susceptible, may result in permanent damage to heart valves. It is common in the Third World, but is now rare in the West (incidence: ~5–10 per million children), although pockets of resurgence have been noted in the USA sometimes in overcrowded areas (favours streptococcal spread). Some specific Group A streptococcal serotypes are known to be particularly rheumatogenic—eg type 5 of the M-protein serotypes. Other ubiquitous serotypes appear to be non-rheumatogenic (eg type 12). medline:191425

Diagnosis The presence of 2 of Jones' major criteria or 1 major and 2 minor **plus** evidence of preceding streptococcal infection: scarlet fever, a throat swab growing β -haemolytic streptococci or a serum ASO titre >333u/L (reference intervals vary). Lancet 2002
medline: 1189728

Major criteria (revised 2001)

Carditis, ie 1 of: changed murmur; ccr; cardiomegaly; friction rub; +ve echo
Polyarthrits (often migratory)
Erythema marginatum, *OHCM* p 416
Subcutaneous nodules
Sydenham's chorea (p 758)

Minor criteria L Veasy 2001 Lancet 357 199

Fever

ESR >20mm or c-reactive protein 1
Arthralgia, ie pain but no swelling
ECG: PR interval >0.2sec
Previous rheumatic fever or rheumatic heart disease

Don't count arthralgia if polyarthrits is a major criterion; likewise for long P-Rs if carditis is used. **Typical joints:** Knees, ankles, elbows & wrists may be very tender, but no permanent sequelae. **Echo criteria:** Mitral regurgitant jet is: >1cm; holsystolic; visible in 2 planes; mosaic pattern (ie chaotic flow).

The MacCullam patch is at the base of the posterior mitral leaflet. Aortic pulmonary and tricuspid valves are affected in descending order of frequency.

Treatment of rheumatic fever Rest/immobilization helps joints & heart

- Aspirin (eg 80mg/kg/day, p 758; but get advice re Reye's syndrome, p 756)
- If severe, get expert help. Prednisolone 2mg/kg/24h may help. medline:1109649
- Penicillin for pharyngitis (eg 125mg/6h po) preceded by one dose of benzylpenicillin (25mg/kg IM or iv).

Prophylaxis Symptoms are likely to be worse on recurrence of rheumatic fever. In one study, recurrence happened in 2.6%. medline:1063812

Prevent this by giving phenoxymethylpenicillin 250mg po daily or sulfadimidine 250mg po daily. Cover surgery/dentistry with antibiotics (see below).

PANDAS (*Paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections*). Suspect this in those with tics (or Tourette's syndrome) and obsessive compulsive disorder. Anorexia nervosa may also be a feature. Antibiotics and risperidone have been tried. medline.10807957 &: 10522469 &: 1093312

Infective endocarditis (IE)

OHCM p 136

Signs Fever, splenomegaly, clubbing, splinter haemorrhages, anaemia, rash, heart failure, microscopic haematuria, new murmur (eg with known congenital heart lesion or iv line *in situ*). **Typical cause:** Streps; staphs. medline:1109655

Tests Blood cultures (different times and different sites), echocardiograms.

Blind treatment Do 3 blood cultures first. Benzylpenicillin 25mg/kg/4h iv + netilmicin 2mg/kg/8h iv (p 238). Get microbiological help, on drugs & duration.

Preventing IE in those with heart lesions Ampicillin 30mg/kg IM/iv + gentamicin 2mg/kg IM at induction, followed by ampicillin 30mg/kg IM/iv 6h later. Metronidazole 7.5mg/kg/8h iv may also be added. This is recommended for any dentistry, colonic or, GU procedure. ▶See *OHCM* p 138.

Some endocrine and metabolic diseases

Hypothyroidism *Congenital:* Thyroid scans divide these into 3 groups: athyreosis; thyroid dysgenesis; and dyshormonogenesis.* Also remember maternal antithyroid drugs (p 157, eg propylthiouracil). *Acquired:* Prematurity; Hashimoto's thyroiditis; hypopituitarism; x-rays; Down syndrome.

Signs: May be none at birth—or prolonged neonatal jaundice, widely opened posterior fontanelle, and dry skin are common features. Inactivity, excessive sleeping, slow feeding, little crying and constipation may occur. Look for coarse dry hair, a flat nasal bridge, a protruding tongue, hypotonia, umbilical hernia, slowly relaxing reflexes, slow pulse and delayed growth and mental development. Other later signs: IQ↓, delayed puberty (occasionally precocious), short stature, delayed dentition. medline:10361481

Universal neonatal screening: Cord blood or filter paper spots (at ~7 days old, from heel prick) allow early diagnosis. They *do* prevent serious sequelae. Act on high and low TSHs (may indicate pituitary failure). medline:10412126

Tests: T₄↓, TSH↑ (but undetectable in secondary hypothyroidism), ¹³¹I uptake↓, Hb↓. Bone age is less than the chronological age. As it is undesirable to x-ray the whole skeleton, the wrist and hand are most commonly used. There are a large number of ossification centres. Each passes through a number of morphological stages, and using comparisons with key diagrams from 'normal' populations, a rough bone age may be determined. There is no hard-and-fast answer to the question of how much discrepancy (eg 2yrs) between skeletal and chronological years is significant.

Using thyroxine: Infants need ~10µg/kg/day (≤50µg/day); this rises to ~100µg daily by 5yrs (adult doses by 12yrs). Adjust according to growth rate and clinical response. *Avoid high TSH levels.* Those with athyreosis need the highest doses of T₄ and the closest monitoring early on. Those with dysgenesis and dyshormonogenesis need more attention later.*

Hyperthyroidism The typical child is a girl at puberty. Clinical and laboratory features are described in *OHCM* p 294. Fine-needle cytology of goitres may reveal *juvenile autoimmune thyroiditis*. medline:11061541

Carbimazole starting dose is eg 250 µg/kg/8h—adjusted according to response. medline:11028118

Propylthiouracil: 2mg/kg/8h po. Expect remission in ~67%. medline:10968475

Thyroid disease in pregnancy and neonates See p157.

The glycogenoses result from defects in synthesis and degradation of glycogen. Abnormal stores are deposited in liver, muscle, heart, or kidney. Most types (there are 7 or so) are inherited as an autosomal recessive. Types include: von Gierke disease (type I, p 759), Pompe's disease (type II, p 756), Cori disease (type III—hypoglycaemia, hepatomegaly, with failure to thrive), Anderson disease (type IV), McArdle disease (type V), Hers disease (type VI) and Tauri disease (type VII—phosphofructokinase↓—like McArdle's, but with haemolysis and gout: diagnose by muscle enzyme assay).

In McArdle's the cause is myophosphorylase deficiency (as shown on muscle biopsy). Inheritance: autosomal recessive. Stiffness and myalgia follow exercise (with electrically silent muscle contractions). During exercise, as circulating fatty acids become available, there may be some recovery ('second wind'). Venous blood from exercised muscle shows low levels of lactate and pyruvate. Lack of phosphorylase staining in muscle biopsy confirms the diagnosis. There may be myoglobinuria (red urine ± renal failure).

Treatment: Avoid extreme exercise. Oral glucose and fructose may help.

*Key reference (2001): A Hanukoglu 2001 *J Clin Endocrinol Metab* **86** 186 [11231999]

born errors of metabolism

These are very numerous, but some helpful generalizations about their acute presentations may be made. Signs include diarrhoea, lethargy, respiratory distress, metabolic acidosis (\pm odd body smells), jaundice, hypoglycaemia, U&E imbalance, fits, and coma. Features may be intermittent, and provoked by crises (eg infection; dehydration). Look for:

<i>Physical sign:</i>	<i>Possible significance:</i>
Hepatosplenomegaly	eg amino acid and organic acid disorders, lysosomal storage diseases (Anderson–Fabry dis.*). Mucopolysaccharidoses, eg Hurler's syndrome, p 750, gangliosidoses, mannosidoses.
Coarse facies	Organic acidurias. See p 204.
Ucc \downarrow , platelets \downarrow	Aminoacidurias, organic aciduria, cystinuria, lactic acidosis, storage diseases.
Mental retardation	
Failure to thrive	

A metabolic urine screen is helpful (details differ from lab to lab—eg amino acids, organic acids, carbohydrates and mucopolysaccharides). Request expert laboratory help. These diseases are associated with an increase in total urinary amino acids:

www.txt.co.uk/full.cfm?1092

Alkaptonuria	Canavan leukodystrophy	Cystinosis
Cystathioninuria	Fructose intolerance	Galactosemia
Hartnup disease	Homocystinuria	Hyperammonemia
Hyperparathyroidism	Maple syrup urine disease	Methylmalonic acidaemia
Osteomalacia	Propionic acidaemia	Rickets
Tyrosinemia type 1 & 2	Viral hepatitis	Wilson's disease

Knowing your way around metabolic disorders (or knowing who to ask) is becoming ever more important, as new molecular techniques provide novel treatment options.¹ Don't be satisfied with genetic counselling and palliative care: find out if this child's condition is treatable.

Anderson–Fabry disease may present with torturing, lancinating pains in the extremities (abdomen) made worse by cold, heat or exercise and not responsive to analgesics. It is due to neuritis caused by vasculitis of the vasa nervorum. By adolescence, angiokeratomata appear (clusters of dark, nonblanching, petechiae) in the 'bathing trunk' area (esp. umbilicus & scrotum). Other signs are acroparaesthesia, corneal opacities, hypohidrosis, proteinuria and renal failure. They may respond to enzyme therapy (α -galactosidase is deficient). Phenytoin or carbamazepine may help the pain.

Acute surgical problems

Congenital hypertrophic pyloric stenosis This does not (contrary to its name) present at birth, but develops during the first month of life ($\sigma/\varnothing \approx 4$).

Presentation: Vomiting which occurs after feeds, and becomes projectile (eg vomiting over far end of cot). Congenital pyloric stenosis is distinguished from other causes of vomiting (eg gastric reflux) by 3 important negatives:

- The vomit does not contain bile, as the obstruction is so high.
- No diarrhoea: constipation is likely (occasionally 'starvation stools').
- Even though the patient is ill, he is rarely obtunded: he is alert, anxious, and always hungry—and possibly malnourished, dehydrated.

Observe left-to-right LUQ (left upper quadrant) peristalsis during a feed (seen in late-presenting babies). Palpate the olive-sized pyloric mass: stand on the baby's left side, palpating with the left hand at the lateral border of the right rectus in the RUQ, during a feed, from a bottle or the left breast. There may be severe water & NaCl deficit, making urine output & plasma Cl^- (also K^+ & HCO_3^-) vital tests to guide resuscitation, and determine when surgery is safe (Cl^- should be $>90\text{mmol/L}$). The picture is of hypochloraemic, hypokalaemic metabolic alkalosis. NB: don't rush to theatre. n=84
medline:11115037

Ultrasound detects early, hard-to-feel pyloric tumours, but is only needed if examination is -ve. ebm
medline:8984923 Barium studies are 'never' needed.

Management: Before surgery (Ramstedt's pyloromyotomy) pass a wide-bore nasogastric tube.

Intussusception The small bowel telescopes, as if it were swallowing itself by invagination. Patients may be any age (eg 5–12mths; $\sigma/\varnothing \approx 3:1$) presenting with episodic intermittent inconsolable crying, with drawing the legs up (colic) \pm vomiting. He may (but need not) pass blood PR (like red-currant jam or merely flecks: do a PR). A sausage-shaped abdominal mass may be felt. He may become shocked, and moribund. **Tests/Management:** The least invasive approach is ultrasound with reduction by air enema (preferred to barium). CT may be problematic, and is less available. Plain abdominal films may be reserved for when perforation is suspected. Doppler studies to show bowel viability have been used but are non-standard. If reduction by enema fails, reduction at laparoscopy or laparotomy is needed. Any necrotic bowel should be resected.

Pre-op care: ►► Resuscitate, crossmatch blood, pass nasogastric tube.

NB: children $>4\text{yrs}$ present differently: rectal bleeding is less common, and they have a long history (eg $>3\text{wks}$) \pm contributing pathology (cystic fibrosis, Henoch-Schönlein or Peutz-Jeghers' syndromes; ascariasis, nephrosis or tumours such as lymphomas—in the latter obstructive symptoms caused by intussusception are the most frequent mode of presentation). Recurrence rate: $\sim 5\%$ (especially in adults, eg after gastrojejunostomy). meta
medline-11178112

Post-operative pain relief Morphine iv: child's loading dose: $100\mu\text{g/kg}$ in 30min, then $10\text{--}50\mu\text{g/kg/h}$ (prem neonates $50\mu\text{g/kg}$, then $5\mu\text{g/kg/h}$)—or diclofenac $0.5\text{--}1.5\text{mg/kg/8--12h}$ if over 1yr (eg a 12.5mg suppository every 8–12h if $\sim 1\text{yr}$ old). Ibuprofen dose: $4\text{--}10\text{mg/kg/6h po}$ (syrup is 100mg/5mL). Pre-op (pre-emptive) tramadol also has advocates ($\sim 0.5\text{--}1\text{mg/kg}$). N=45
medline:10930636

Phimosis (The foreskin is too tight, eg due to circumferential scarring, so that retraction over the glans is impossible, eg with foreskin ballooning on micturition \pm balanitis.) It is normal to have a simple non-retractile foreskin up to the age of 4yrs. Time, or a wait-and-see policy will usually obviate the need for circumcision. Forced retraction may be causative, not therapeutic. 0.05% betamethasone cream is also reported to be effective. n=23
medline:10458456

Other surgical problems: **appendicitis, hernias, volvulus, torsion of the testis, acute abdomen:** see p 272, p 218, OHCM p 482 and OHCM, p 462.

Precocious puberty

Puberty may start as early as 8yrs of age in girls and 9yrs of age in boys. Refer to a paediatric endocrinologist if onset is before this. $\text{♀}/\text{♂}=4:1$. medline:11133068

Biology Each of the physical signs of puberty may be thought of as a bioassay for a separate endocrine event. Enlargement of the testes is the first sign of puberty in boys, and is due to pulses of pituitary gonadotrophin. Breast enlargement in girls and penis enlargement in boys is due to gonadal sex steroid secretion. Pubic hair is a manifestation of adrenal androgen production. The growth of boys accelerates when the volume of the testis reaches 10–12mL (as measured by comparison with the orchidometer's beads). Girls start to grow more quickly once their breasts have started to develop. Stage 4 breast development is a prerequisite for menarche (in most girls). The first clue to distinguish simple from pathological precocity is when this *consonance of puberty* goes awry: in Cushing's syndrome, pubic hair is 'too much' for the testicular volume; in hypothyroidism, the testes are large ($\text{FSH}\uparrow$ because $\text{TSH}\uparrow\uparrow$) in the presence of a low growth velocity. medline:8984908

Signs of precocious puberty (variable) boys: rapid growth of penis and testes, increasing frequency of erections, masturbation, appearance of pubic hair, changing body odour, and acne. There will be corresponding changes in the secondary sexual characteristics of girls. The chief consequence is short stature caused by early fusion of epiphyses. Premature adrenal maturity (adrenarche) may presage insulin resistance & syndrome X. medline:11133068
Enquire about symptoms of more general hypothalamic dysfunction: polyuria, polydipsia, obesity, sleep, and temperature regulation. There may also be manifestations of raised intracranial pressure and visual disturbance.

In girls, often no cause is found, but in 85% of boys a cause is found. If onset is before 2yrs, the cause may well be a hamartoma in the hypothalamus—seen on MRI/CT. LH receptor gene mutations are the cause of sporadic or familial male gonadotrophin-independent precocious puberty. medline:11134146
Genetic mechanisms include substitution of τ for c at nucleotide 1103, resulting in substitution of leucine at position 368 by proline. *Other causes:*

- CNS tumours, hydrocephalus, or craniopharyngioma
- Tuberous sclerosis
- Post encephalitis, meningitis, or cranial irradiation
- Hepatoblastoma
- McCune-Albright syn. (\uparrow oestrogen from ovarian cysts)
- Choriocarcinoma
- Primary empty sella syndrome medline:7645385
- Hypothyroidism

Tests Cranial CT/MRI; bone age by skeletal x-ray; urinary 17-ketosteroids; karyotype analysis; pelvic ultrasound (girls), T_4/TSH . medline:10969912 & 10821227

Management: a physiological approach Initiation of puberty depends on release from inhibition of neurones in the medial basal hypothalamus that secrete gonadotrophin-releasing hormone (GnRH), and on decreasing hypothalamic-pituitary sensitivity to $-ve$ feedback from gonadal steroids. These changes are accompanied by a marked increase in frequency and magnitude of 'pulses' of luteinizing hormone (LH), and, to a lesser extent, follicle stimulating hormone (FSH). It is the ability to secrete GnRH pulses at a fast rate which leads to normal gonadal function. Continuous high levels of GnRH paradoxically suppress the secretion of pituitary gonadotrophins, and this forms the basis for treatment of precocious puberty with synthetic analogues of GnRH . After subcutaneous or nasal insufflation there is a reversal of gonadal maturation and all the clinical correlates of puberty (not for pubic hair, as there is no change in the secretion of androgens by the adrenal cortex). There is deceleration in skeletal maturation. Treatment is continued eg up to 11yrs old. Families need reassurance that the child will develop normally.

Endogenous oestrogen accelerates growth (♂ & ♀), as well as mediating ♀ sexual characteristics: testolactone can help by \downarrow its biosynthesis. Androgens (eg flutamide) and spironolactone also have a rôle. medline:10731123

Primary antibody deficiency

Essence Synonyms for this condition include primary hypogammaglobulinaemia, ie not secondary to protein-losing enteropathy, chronic lymphatic leukaemia, or myeloma. Prevalence: ~4/100,000. The leading symptom is recurrent infections, needing repeated antibiotics, with the development of chronic infective states such as bronchiectasis and sinusitis. Other features:

- Failure to thrive
- Nodular lymphoid hyperplasia (gut)
- Absent tonsils
- Enteropathy
- Hepatosplenomegaly
- Anaemia

Most diagnoses are in children >6yrs (presentation may also be in adulthood).

Types of primary immunodeficiency IgA deficiency (39%); common variable immunodeficiency (14%; IgG↓, IgA↓, IgM variable), severe combined immunodeficiency/T-cell defects (13%), complement deficiency (10%), X-linked agammaglobulinaemia (4%; often ♂; B cells may be abnormal), IgG subclass deficiency (3%), chronic granulomatous disease (3%). medline:11434886

Suspect immunodeficiency if

- Failure to thrive
- Recurrent infections
- Severe infections needing surgery
- Arthropathy
- Hepatosplenomegaly

Lab clues: lymphopenia; ↔ serum albumin but ↓total protein (immunoglobulins are missing, so making crossmatching hard, as no haemagglutinins). medline:9033500

When these signs are unexplained, refer to an immunologist, to assess antibody responses to protein and carbohydrate antigens, measure IgG subclasses, and count lymphocytes involved in antibody production (CD4, CD8, CD19, CD23 positive lymphocytes). Immunoglobulin levels are interpreted by age. There is a rôle for watching responses to test vaccinations.

Management Aim to include the patient and the family in the process. Ensure prompt treatment of intercurrent infections. This may include postural physiotherapy, and bronchodilators as well as antibiotics.

Immunoglobulin replacement obviates most complications and is best delivered by a specialist immunologist, after detailed immunological assessment. Many patients can join a self-infusion programme. Before any infusion, active infection should be excluded (to minimize risk of adverse reactions), and a baseline check of transaminase enzymes, creatinine, and anti-IgA antibody titres should be done.

The dose of iv immunoglobulin is determined by the severity and frequency of infections, and the plasma level of IgG. Most receive ~400mg/kg/month, usually as 2 doses, 2 weeks apart. medline:8148684

Have hydrocortisone and an antihistamine at the ready. SE: headaches, abdominal pain, anaphylaxis, transmission of hepatitis. medline:11131000

Intramuscular immunoglobulins are not favoured, but the subcutaneous route is being investigated and appears satisfactory. N=40 medline:10821460

Complications *Chest:* Bronchiectasis, granulomas, lymphoma.

Gut: Malabsorption, giardia, cholangitis, atrophic gastritis, colitis.

Liver: Acquired hepatitis, chronic active hepatitis, biliary cirrhosis.

Blood: Autoimmune haemolysis, ITP, anaemia of chronic disease, aplasia.

Eyes/cns: Keratoconjunctivitis, uveitis, granulomas, encephalitis.

Others: Septic arthropathy, arthralgia, splenomegaly.

Gene therapy* Autologous haematopoietic stem cells transduced with the γ c gene can restore immune system in boys with severe combined immunodeficiency. A harmless retrovirus carries the replacement gene, and infects the stem cells *in vitro*. When these are replaced in the marrow an immune system develops within a few months[†]—obviating the need for intrusive anti-infection isolation measures and iv Ig. It is an alternative to marrow transplants.

© C Hue 2002 *NEJM* 346 1185 n=5. [†] T cells & repertoires of T-cell receptors were ~normal up to 2yrs post-op; thymopoiesis is shown by naïve T cells. Antibody production is adequate.

Delay in walking

Babies usually learn to walk at ~14 months old. If this has not occurred by 20 months, ask yourself 2 questions: Is the child physically normal? Is development delayed in other areas too? The commonest causes reflect chronic illness, global delay, benign immaturity, and generalized joint hypermobility. In boys consider Duchenne muscular dystrophy early (genetic counselling before mother's next pregnancy). Note that congenital hip dislocation may present as 'walking delay' if the fact of limping is overlooked. The major reason for identifying late walkers is to exclude cerebral palsy.

Cerebral palsy comprises a range of chronic disorders of posture and movement caused by a non-progressive CNS lesion sustained before 2yrs old, resulting in delayed motor development, evolving CNS signs, \pm learning disability (35%) \pm epilepsy. Most are due to antenatal events unrelated to birth trauma. *Prevalence*: 2/1000 (50% are of low birth weight). *Typical survival*: 20yrs if quadriplegic (much longer if less affected). $\text{♀}/\text{♂}=1:1$. medline:10537930

Prenatal factors:

APH (with hypoxia)
X-rays; alcohol
CMV; rubella; HIV
Toxoplasmosis
Rhesus disease

Perinatal factors:

Birth trauma
Fetal distress
Hypoglycaemia
Hyperbilirubinaemia
Resuscitation problems

Postnatal factors:

Trauma; Intraventricular haemorrhage
Meningoencephalitis
Cerebral vein thrombosis (from dehydration)

Clinical picture: Weakness, paralysis, delayed milestones, seizures, language and speech problems. Spasticity suggests a pyramidal lesion; unco-ordinated, involuntary movements and postures (dystonias) suggest basal ganglia involvement. Most children have either a hemiparesis or a spastic diplegia—eg both legs affected worse than the arms, so that the child looks normal until he is picked up, when the legs 'scissor' (hip flexion, adduction and internal rotation; with knee extension and feet plantar-flexed).

Type 1 Ataxic palsies (pure ataxia):

Hypotonia ('floppy baby')
Other defects rare
Flexor plantars
Associations: deafness; strabismus
mental retardation (fits rare)
Normal developmental milestones

Type 2 Ataxic diplegia:

Hypertonia
Other defects common
Extensor plantars
Associations: trauma
hydrocephalus
spina bifida; viruses

Dyskinetic cerebral palsy: Unwanted actions, poor movement flow, agonist/antagonist imbalance, poor posture control, hypotonia, hearing \downarrow , dysarthria.

Management This is multidisciplinary (include physiotherapists, occupational therapists, orthopaedic surgeons, and orthoses experts). Assess function: can he roll over (both ways)? Can he sit? Grasp? Transfer objects from hand to hand? Good head righting? Ability to shift weight (when prone) with forearm support. IQ. Is toileting possible? Can he hold a pen or a spoon? Treat epilepsy (p 268). Calipers may prevent deformity (equinovarus, equinovagis, hip dislocation from excessive flexion/adduction). Attempts to show benefits of neurodevelopmental physiotherapy (to improve equilibrium and righting) don't show benefit over simple stimulation of motor activity. Some parents try the Hungarian *Pető approach*: here the 'conductor' devotes herself to the child and uses interaction with peers to reinforce successes: manipulation, art, writing, fine movement, social skills. medline:2513029

Botulinum toxin (p 522) has an uncertain rôle. meta-analysis medline:10796784 & 11851730 & 11087280

Epidural cord electrostimulation and intrathecal or oral baclofen: These are novel therapies needing further evaluation. medline:11221337 & 9285607

Prognosis By 6yrs, 54% with quadriplegia (80% if hemiplegic or diplegic) gain urinary continence spontaneously. If IQ \downarrow , 38% are dry at this age. n=601 medline:11221896

Delay in talking

►When in doubt, ask the mother; test the hearing. *Ensure the result is reliable.*

Speech development After the first few months of life, language differentiation is already occurring. Chinese babbling babies can be distinguished from European ones. Before the first year babbling gives way to jargon—plausible-sounding ‘words’ which have little by way of fixed meaning.

- At ~1 year . . . a few words may be used meaningfully.
- At 1½ yrs old. . . 2-word utterances appear (“Daddy come”).
- At 2 years old. . . subject–verb–object sentences appear (“I want a pudding”).
- At 3½ yrs old. . . the child has mastered thought, language, abstraction, and the elements of reason, having a 1000-word vocabulary at his or her disposal, enabling sentences such as: “I give her cake ‘cos she’s hungry”.

Words exist to give ideas currency, and so often that currency proves counterfeit—a process which so often starts with *if*, eg “*If I hadn’t thrown the cup on the ground, I might have got a pudding*”. The uttering of “*If. . .*”, linked with an emotional response, is the most human of all constructions, opening up worlds divorced from reality, providing for the exercise of imagination, the validation of dreams, the understanding of motives, and the control of events. The rest of life holds nothing to match the intellectual and linguistic pace of these first years. Further linguistic development is devoted to seemingly conceptually minor tasks, such as learning the subjunctive, and expanding vocabulary.

There is much variation in speech timing: what is clearly abnormal?

Vocabulary size: If <50 words at 3yrs old, suspect deafness—or:

- Expressive dysphasia or speech dyspraxia (eg if there is a telegraphic quality to speech, poor clarity, and deteriorating behaviour, eg frustration).
- Audio-premotor syndrome (APM). The child cannot reflect the sounds correctly heard into motor control of larynx and respiration. Instead of babbling, the child is quiet, unable to hum or sing.
- Respiro-laryngeal (RL) dysfunction (dysphonia from incorrect vocal fold vibration/air flow regulation). The voice is loud and rough.
- Congenital aphonia (rare): the voice is thin, weak, and effortful.

Speech clarity: By 2½ years old, parents should understand their child’s speech most of the time. If not, suspect deafness—or:

- Articulatory dyspraxia (easy consonants are *b* and *m* with the lips, and *d* with the tongue—the phonetic components of babbling). ♂: ♀ ≈ 3:1. Tongue-tie is a possible cause, and they will have trouble with sounds needing tongue elevation (*d* and *s*)—surgery to the frenum may be needed as well as speech therapy. Distinguish from phonological disorders (disordered ‘sound for speech processing’—may present as ‘sound awareness’ problems (ie difficulty in analysing the sound structure of words). Both are common.
- APM or RL dysfunction, as described above.

Understanding: By 2½ years he should understand “Get your shoes”, if not suspect: •Deafness—if the hearing is impaired (eg 25–40dB loss) secretory otitis media is likely to be the cause. Worse hearing loss is probably sensorineural •Cognitive impairment •Deprivation.

Some other causes of speech disorder For autism, see p 392. Others:

Acquired:	Congenital:
Post meningoencephalitis	Klinefelter’s syndrome
Post head injury	Galactosaemia, histidinaemia
Landau–Kleffner syndrome	Auditory agnosia
(epilepsy + progressive loss of language)	Floating-harbo(u)r syndrome*

Speech therapy Refer early, before school starts. NB: randomized trials have not shown any clear benefits from this strategy.

N=159
medline~11030677

Named after a boy seen at the Boston Floating Hospital with short stature + delayed bone age + expressive language disorder, + triangular face with prominent nose, hyperkinesis, coeliac disease, short hands.

Impairment, disability, and handicap

WHO definitions Impairment entails a pathological process, eg spina bifida, which may cause certain disabilities, eg walking difficulty. Handicap is their social consequence (eg cannot walk to school).

Learning delay (mental handicap) The mother often makes the first diagnosis. An IQ <35 constitutes a severe learning disability (mental handicap, p 314).

►Beware conflating IQ with intellect: the latter implies more than problem-solving and memory: intellect entails the ability to speculate, to learn from mistakes, to have a view of oneself and others, to see relationships between events in different domains of experience—as well as the ability to use language either to map the world, or to weave ironic webs of deceit (and, on a good day, to do both simultaneously).

Causes: Severe mental impairment usually has a definable cause, whereas mild retardation is often familial, with no well-defined cause. Be prepared to refer to an expert. **Congenital disorders** are legion: chromosomal (eg Down's; fragile X, p 752); metabolic (eg PKU p 205). **Acquired:** Perinatal infection p 98–100, birth injury and cerebral palsy, trauma, meningitis.

Lead exposure: This is a leading preventable cause of mildly impaired intellect. For example in 2-yr-olds for each 0.48 μmol/L plasma increment there is an associated 5–8 point fall in IQ as measured on the Wechsler Intelligence Scale for children (revised). This defect is long-lasting.

Biochemical defects associated with mental retardation—eg:

Homocystinuria: Paraplegia, fits, friable hair, emboli, cataracts; homocystine is found in the urine. Treat with a low methionine, cystine-supplemented diet, with large doses of pyridoxine.

Maple syrup urine disease: Hypoglycaemia, acidosis, fits, death. Urine smells of maple syrup, due to defective metabolism of branched chain keto acids. Treatment: high calorie amino-acid controlled diet. Thiamine has been tried.

Tryptophanuria: Rough, pigmented skin. Treat with nicotinic acid.

Management: Refer to an expert, so that no treatable cause is missed. Would the family like help from a self-help group, such as MENCAP? Other members of the family may need special support (eg normal siblings, who now feel neglected). If the IQ is >35, life in the community is the aim.

Physical handicap **Sensory:** Deafness, see p 540.

Blindness: congenital defects are described on p 516. Principal acquired causes of blindness are: retinopathy of prematurity, vitamin A deficiency, onchocerciasis (p 512), eye injuries, cataract (eg Down syndrome).

CNS & musculoskeletal problems: (Congenital or acquired) Causes: accidents (eg near-drowning), cerebral palsy (p 202), spina bifida (p 228), after meningitis, polio, congenital infections (above), tumours, syndromes (p 742–758).

Wheelchairs:³ For indoors or outdoors? Patient-operated, motorized or pushed? What sort of restraints to prevent falling out? If collapsible, how small must it be to get into the car? Are the sides removable to aid transfer from chair to bed? Can the child control the brakes? Are there adjustable elevated leg rests? Liaise with the physio and occupational therapist.

Calipers will allow some patients to stand and walk. Long-leg calipers are required for those with complete leg paralysis. The top should be constructed so that it does not induce pressure sores. A knee lock supports the knee in the standing position. An internal coil spring prevents foot drop.

³Disabled Living Foundation, 380 Harrow Rd, London W9, UK tel 027289 6111

Check-list to guide management of handicap

Whether based in hospital or in the community the doctor should address each of these points:

- Screening and its documentation on local handicap registers.
- Communication with parents.
- Refer to and liaise with district handicap teams, and community paediatrician.
- Ensuring access to specialist services, including physiotherapy, orthopaedic surgery.
- Assessing special needs for schooling and housing.
- Co-ordinating neuropsychological/neurodevelopmental assessments.
- Co-ordinating measures of severity (eg electrophysiology \pm CT/MRI).
- Liaison with dietitian on special foods.
- Promotion of long-term compliance with treatment or education programmes.
- Education about the consequences of the illness.
- Encourage contact with family support groups.
- Offering family planning *before* patients become unintentionally pregnant.
- Pre-conception counselling (with specialist in molecular genetics).
- Co-ordinating prenatal diagnostic tests and fetal assessment.

Phenylketonuria (PKU)

Cause Absent or reduced activity of phenylalanine hydroxylase—inherited as an autosomal recessive (the gene—on chromosome 12—often mutates, and there is a full range of clinical manifestation from nearly symptom-free to severe mental impairment). The defect leads to \downarrow CNS dopamine, reduced protein synthesis, and demyelination. medline:11043156

Clinical features Fair hair, fits, eczema, musty urine. The chief manifestations is \downarrow IQ (eg dyscalculia \pm poor spelling \pm \downarrow cognition). medline:11138905

Tests Hyperphenylalaninaemia (reference interval: 50–120 μ mol/L). 'Benign' phenylketonuria may be indicated by levels $< 1000 \mu$ mol/L.

Treatment See check-list above. *Diet:* protein substitute that lacks phenylalanine but is enriched in tyrosine. Aim to keep phenylalanine to $< 600 \mu$ mol/L—by prescribing artificial food-substitutes (amino acid drinks) to give < 300 mg–8g of natural protein/day (depending on age and severity of phenylalanine hydroxylase deficiency). medline:11157309

There is evidence to suggest that hypomyelination is proportional to degree of phenylketonaemia, but some studies fail to show stricter diets are associated with higher IQs. Despite treatment, children are more prone to depression, anxiety, phobic tendencies, isolation, and a less 'masculine' self image. In one meta-analysis, features of hyperactivity disorder were documented, as well as associations between behaviour problems and both lower IQ and a worse quality of PKU control. medline:11043152

Adherence to the diet may be poor (it's unpalatable). medline:11043160
Also, the diet may cause changes of questionable significance in levels of selenium, zinc, iron, retinol, and polyunsaturated fatty acids. medline:11043159

Prevention of manifestations Screen blood at 1 week (using a heel-prick and filter paper impregnation—the Guthrie test).

Maternal phenylketonuria However good the mother's control, subtle neurological impairment is likely. The implication is that control during pregnancy should be even tighter than in childhood. Hence the need for pre-conception counselling.

Behavioural problems

► *Only enter battles you can win.* If the child can win, be more subtle (eg consistent rewards, not inconsistent punishments). Get health visitor's advice, and ensure that everyone is encouraging the same response from the child.

Entrances *Food refusal* and *food fads* are common. Reducing pressure on the child, discouraging parental over-reaction, and gradual enlarging of tiny portions of attractive food is usually all that is needed. Check Hb. *Overeating*: Eating comforts, and if the child is short on comfort, or if mother feels inadequate, the scene is set for overeating and life-long patterns are begun. Diets may fail until the child is hospitalized (p 348).

If obese, remember hypothalamic syndromes (eg Prader–Willi, p 756).

Pica is eating of things which are not food; all will do this at some time; if it is persistent, there may be other signs of disturbed behaviour, or ↓ IQ. Some evidence suggests that pica is best classified as an obsessive-compulsive disorder. *Complications*: lead poisoning; worm infestations. medline:11001006

Exits *Soiling* is the escape of stool into the underclothing.

Encopresis is the repeated passage of solid faeces in the wrong place in those >4yrs old. Faecal retention is the central event. *Constipation* is difficulty in defecation; it may lead to distress, abdominal pain, rectal bleeding, abdominal masses, overflow soiling ± 'lavatory-blocking' enormous stools (megarectum), and anorexia. Causes: often poor food, fluid, or fibre intake. Fear—eg as a result of a fissure. Rarely Hirschsprung's disease (think of this in infants if there is an explosive gush of faeces on withdrawing the finger after rectal exam, or if there is alternating diarrhoea with constipation and abdominal distension, and failure to thrive). *Action*: • Find out about pot refusal • Does defecation hurt? • Is there parental coercion? Break the vicious cycle of: large faeces → pain/fissure → fear of the pot → rectum overstretched → call-to-stool sensation dulled → soiling → parental exasperation → coercion). Exonerate the child to boost confidence for the main task of obeying the call-to-stool to keep the rectum empty (aided by *docusate sodium* (both a softener and a stimulant), then a dose of *sodium picosulfate* elixir or *polyethylene glycol*, followed by maintenance *methyl cellulose tablets* ± prolonged alternate-day *senna*). Psychological methods (eg biofeedback) have *not* proved efficacious. meta analysis
medline:11211851

Give the family time to air feelings which encopresis engenders (anger, shame, ridicule). Treat perianal cellulitis with co-amoxiclav.

Enuresis: Bedwetting occurs on most nights in 15% of 5-yr-olds, and is still a problem in up to 3% of 15-yr-olds—usually from delayed maturation of bladder control (there is often a +ve family history). Tests for diabetes, UTI and GU abnormality (p 188) can occasionally yield surprises. The term 'secondary enuresis' implies wetness after >6 months' dryness. *Treatment*: Avoiding acrimonious blame, fluid restricting before bed, and the passing of time may help. Reassure the child that he is neither infantile nor dirty. A system of rewards for dry nights as documented on a star chart above the child's bed may be effective. Alarms (± vibrations) triggered by urine in the bed can make 56% dry at 1yr; relapse rate are preventable by continuing use after dryness. They are cheap or loanable from the Community Health/Child Guidance Services (or equivalent)—eg Drinite®. www.bedwetting.co.uk Imipramine 25–50mg/24h po should not be used for >3 months, has side-effects, and high relapse rates on discontinuation. Desmopressin (DDAVP) 20–40µg (2–4 nasal puffs/night) helps short-term. Oral dose (if >5yrs): 0.2mg at bedtime. Beware fluid overload, eg after gulping in swimming pool water, or going to rave parties; have 1 week 3-monthly with no drugs. ci: cystic fibrosis, uraemia, BP↑. Oxybutynin 2.5–5mg po at night may help if bladder irritability. meta analysis
medline:10908533 & 10479880

Hyperactivity/ADD The spectrum ranges from a level of activity > parents are happy with, to serious lack of concentration, impulsivity, and scholastic failure—known as *attention deficit disorder (ADD)* or *minimal brain dysfunction*. ADD is commoner in learning-disabled children, and if there has been prenatal exposure to cannabis.

2001 medline:11274871 & 10840176

Parents say the child is never still, being 'on the go, as if driven by a motor', ± :
 Squirming and fidgeting Inconsiderate of others Blurts out answers
 Sustained attention ↓ Interrupts continually Behind with schoolwork
 Speech ↑ Instructions never obeyed Butts in during games

Not all those with ADD are hyperactive. *There is no diagnostic test* (but positron emission tomography may show ↓ function of frontal lobes and near connections). Most parents first note excessive activity at the toddler stage, but most locomotor hyperactivity at this stage abates with time, so the diagnosis is usually delayed until school entry or later. Signs generally attenuate during adolescence, but may persist into mid-adulthood. There is a familial tendency, and families with ADD are more likely to have other diagnoses too, such as learning difficulties, mood, anxiety, and psychosocial disorders (DSM IV, p 312). *Treatment:* Take time to explain the condition. Offer a booklet. Cognitive therapy may increase parental tolerance. Hypoallergenic diets (eg no tartrazine) are controversial and may cause obsessions in parents.

Psychostimulants (not depressants) can help (confer with an expert): methylphenidate 5mg/12–24h po (max 30mg/12h) is said to offer the best benefit-to-hazard ratio. SE: insomnia, slowing of growth; monitor growth, BP, pulse. Many of the side-effects associated with stimulant use appear, to be relatively mild and of short duration, and to respond to dosing or timing adjustments. Evidence supporting long term use is very uncertain. ^{meta-analysis} medline:10790990 & 11762571

Other drugs: pemoline 0.5–2mg/kg/24h po (effects are delayed for 6 weeks); haloperidol 0.01–0.04mg/kg/12h po; imipramine 0.2–0.5mg/kg/ 8h, especially if co-existing anxiety or depression; clonidine (≤5mg/day po in divided doses, particularly if tic disorder is also present). medline:10660814

School refusal *Setting:* Emotional overprotection; high social class; neurotic parents; schoolwork of high standard. In truancy, the reverse is true. *Treatment:* Confer with headteacher, parents, and an educational psychologist. Escort by an education welfare officer aids the prompt return to school. Often anxiety and/or depression will also need treatment. medline:11211369

Screening and child health promotion

The main aims: ●Encouraging breast feeding (p 176–8) ●Monitoring development ●Immunization ●Overseeing growth (p 184) ●Parental support ●Education and reassurance about normal childhood events. ●Talking to the child, and building up a good relationship to be used in later illnesses.

Monitoring The most cost-effective times to screen are unknown. A 'best buy' might be checks after birth (p 231), at 4–6 weeks—arrange vaccinations and $\text{D}_{0-6\text{H}}$ vitamin drops* (A, D & c, unless sure that diet/sunlight is adequate), then:

6–9 months: Hips, testes descent, cvs examination.

18–24 months: Educate on diet, dental care, accident; walking (look for waddling), social and linguistic milestones; Hb if iron deficiency likely—it may well be. Any parental depression?

4 years: Testes descent, cvs examination. Nutrition, dental care.

At each visit: ●Encourage airing of queries. ●Ask about, and test for, squint, vision and deafness.† ●Chart centiles. Beware reading too much into a single test. See Denver developmental test, p 288. Remember to correct age for prematurity. Note the milestones below. There is much individual variation.

1 month: Lifts head when prone; responds to bell; regards face.

2 months: Holds head at 45° when prone; vocalizes; smiles.

4 months: Uses arm support when prone; holds head steady when supported while sitting; reaches out; spontaneous smiling.

6 months: Bears some weight on legs; on pulling to sitting, there is no head lag; reaching out; transfers things from hand to hand.

≥ 1 year: Just stands; walks using a table's support; clashes cubes; pincer grip; can say 'Mummy' \pm 'Daddy'. Plays 'pat a cake'.

18 months: Can walk backwards; scribbles; 2-cube tower. 2–4 words.

Recognizes/shows interest in tv images, eg of a dog.[‡]
NB: drooling \pm throwing items on the floor is now abnormal.

2 years: Kicks a ball; overarm 'bowling'; gets undressed.

3 years: Jumps; can stand on one foot; copies; can build an 8-cube tower; knows his first and last name; dressing needs help.

4 years: Stands on 1 foot for $>4\text{sec}$; picks the longer of 2 lines.

Immunization Offer the schedule given OPPOSITE, noting the items below.

MMR vaccine:* There is no upper age limit. **se:** Fever from day 5 (so give antipyretic advice); rare non-infectious parotid swelling, from week 3. **ci:** Fever, pregnancy (advise against this for 1 month), a previous live vaccine within 3 weeks or immunoglobulin within 3 months, primary immunodeficiency syndromes (not including HIV or AIDS), those receiving prednisolone $\geq 2\text{mg/kg/day}$ for >1 week in the last 3 months, leukaemia, lymphoma, recent radiotherapy, anaphylaxis induced by egg, neomycin, or kanamycin (vaccine preservative). Non-anaphylactoid allergies are not contraindications, and nor is a past history of seizures or febrile convulsions.

Pertussis immunization se: Pain and fever. Serious brain damage is so rare, that if it occurs at all, it is very hard to put a figure to. It is equivocally implicated in 1 in 2×10^6 injections. **ci:** Past severe reaction to pertussis vaccine—ie indurated redness most of the way around the arm, or over most of the anterolateral thigh (depending on the site of the injection); or generalized reactions: fever $>39.5^\circ\text{C}$ within 48h of vaccination; anaphylaxis; prolonged inconsolable screaming; or other CNS signs (eg fits) occurring within 72h.

Special considerations: Most children with idiopathic epilepsy, or a family history of epilepsy (sibling or parent) should be vaccinated. If in doubt, seek expert advice rather than withholding the vaccine. Those with stable CNS conditions (eg cerebral palsy, spina bifida) are especially recommended for vaccination. There is no upper age limit. Collapse (sudden loss of muscle tone, pallor, and unresponsiveness) is rare, and subsequent doses may be given without complication. A single-antigen acellular vaccine is available (tel: ^{UK} 01708 379000).

Vaccination schedules

- ▶ An acute febrile illness is a contraindication to any vaccine. Note:
- Give live vaccines either together, or separated by ≥ 3 weeks.
- Don't give live vaccines if there is a primary immunodeficiency, or if taking steroids (eg $>2\text{mg/kg/day}$ of prednisolone); but if HIV+ve, give all immunizations (including live) except for BCG, in areas where TB prevalence is low.

Age of child: Immunization (L = live vaccine; MCC=meningitis C conjugate vaccine)

3 days BCG^L (if TB in family in last 6 months or the family comes from area of high TB prevalence); avoid in some immunodeficiencies.[†]

2 months DTWP-Hib (diphtheria, tetanus, whole cell pertussis, haemophilus influenzae type b, eg in a ready-mixed vial; if DTWP is used as diluent for Hib, shake vigorously); and MCC (meningitis C conjugate vaccine) and oral Polio^L. If premature, still give at 2 months. Acellular pertussis (DTaP-Hib; DTaP-IPV/Hib) is non-standard at this age in UK (?fewer SEs, but less immunogenic and more expensive[•]) but DTaP is standard eg in Germany. medline:11734739

3 months Repeat DTWP-Hib and MCC and Polio^L.

4 months Repeat DTWP-Hib and MCC and Polio^L.

12-18 mths 1st dose of Measles/Mumps/Rubella^L (=MMR-II; 0.5mL sc).

4-5yrs DTaP (diphtheria, tetanus and acellular pertussis) medline:10600188 and Polio^L booster; dose 2 of MMR-II.^{**}

10-14yrs BCG^L. (Also Rubella^L for girls who have missed MMR-II.)

15-18+yrs Polio^L and Tetanus and low-dose Diphtheria Td boosters; MCC if unvaccinated and aged between 15 and ~24yrs.

Any age One-off 23-valent pneumococcal vaccine + yearly 'flu vaccine if chronic heart, chest, or renal disease; DM; immunosuppression (eg HIV +ve, cirrhosis, on chemotherapy, or spleen function \downarrow , eg related to coeliac or sickle cell diseases). Consider a 2nd pneumococcal vaccine if at \uparrow risk after ~5yrs. medline:11745926
Some recommend[•] 3 doses of pneumococcal conjugate vaccine if HIV+ve (separated by 2mths, if <2 yrs old). medline:9093301 & 11564590

Adults Boosters eg of Tetanus and Polio. Travellers: see OHCM p 560.

Hepatitis B (Engerix B[®]): Give at birth, 1 & 2 months, if mother is HBsAg +ve 0.5mL IM via anterolateral thigh (adult dose: 1mL via deltoid; 1st dose accompanied by 200u hepatitis B immunoglobulin at a different IM site, eg if high risk, ie e antibody -ve or e status unknown). Other risk groups: OHCM p 230.

Immunization in immunodeficiencies: See Royal College^{paeds} Guidelines.[†]

[†]2002 Immunization of the Immunocompromized Child. www.rcpch.ac.uk [Immunocomp pdf]

[†]A recent meta-analysis suggests that there is no value in these hearing tests: BJGP 1999 49 4

[•]B Wharton 1999 BMJ i 2 (get from Health Visitor); dose of vit. D: 7 $\mu\text{g/day}$; iron may also be needed (also 7 $\mu\text{g/day}$). See also J Labadie 1998 BMJ i 902 & D Hall 1995 Health for all Children, 3e and UK DoH

^{**}The aim of the 2nd MMR dose is to eliminate measles (as in Finland). If we relied on a single dose, herd immunity would \downarrow , and every 5yrs an epidemic could occur, as in the USA in 1990, when 135 babies died (10% were older children who had been vaccinated). We are aware of tenuous reports of associations between vaccination and bowel disease \pm autism.[•] This has been fully considered by vaccination committees in many countries—none has changed its advice. B Taylor 2002 BMJ i 393

Genetic disease and prevention

► It is more important to be able to love the handicapped and to respect their carers than it is to prevent the handicap—for in doing the first we become more human. In the second, we risk our own inappropriate deification.

Gene probes These use recombinant DNA technology to link genetic diseases of unknown cause to DNA markers scattered throughout the human genome. Using fetal DNA from amniotic fluid cells (amniocentesis, p 166) in the second trimester, or from chorionic villus sampling in the first, it is possible to screen for many genetic diseases (opposite)—eg Huntington's chorea; muscular dystrophy; polycystic kidneys; cystic fibrosis; thalassaemias.

Enzyme defects Many of the inborn errors of metabolism can be diagnosed by incubation of fetal tissue with a specific substrate.

Chromosomal studies can be undertaken on cultured cells or on direct villus preparations. The most important abnormalities are aneuploidies (abnormalities in chromosome number)—eg trisomy 21, 18, and 13.

Screening for chromosomal abnormalities (eg the fragile X syndrome, p 752) may be performed on at-risk mothers who may be carriers.

Nondisjunction After meiosis one gamete contains two number 21 (say) chromosomes* and the other gamete has no 21 chromosome. After union of the first gamete with a normal gamete the conceptus has trisomy 21, and develops Down syndrome (but 50% of these will spontaneously abort). This is the cause in $\geq 88\%$ of Down's babies, and is related to maternal age (the mechanism is unknown). *Risk at 20yrs: 1 in 2000; 30yrs: 1 in 900; 35yrs: 1 in 365; 40yrs: 1 in 110; 45yrs: 1 in 30; 47yrs: 1 in 20.* medline:11173856 & 10935320

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Swapping chromosomal fragments Cells contain the correct amount of genetic material (a balanced translocation); but any gamete cell may have one intact chromosome (say number 21), and another mixed chromosome (say number 14), which, because of the swap, contains material from chromosome 21. After union with a normal gamete, cells have 3 parts of chromosome 21. This translocation trisomy 21 is the cause in 4% of Down syndrome (unrelated to maternal age). If the father carries the translocation, risk of Down's is 10%; if it is the mother, the risk is 50%. 0.3% of mothers have this translocation.

Mosaicism A trisomy may develop during early divisions of a normal conceptus. If the proportion of trisomy 21 cells is low (eg 4%) cognitive development may be normal. It accounts for $\leq 8\%$ of Down's babies. medline:10711655

Other chromosomal abnormalities Turner's (p 759), Klinefelter's (p 750), Edward's (p 746), and Patau's (p 754) syndromes. In the *cri-du-chat* syndrome there is deletion of the short arm of chromosome 5, causing a high-pitched cry, cvs abnormalities, microcephaly, widely-spaced eyes, and a 'moon' face.

Down syndrome *Causes:* See above. *Recognition at birth:* Flat facial profile, abundant neck skin, dysplastic ears, muscle hypotonia, and x-ray evidence of a dysplastic pelvis are the most constant features. Other features: a round head, protruding tongue, peripheral silver iris spots (Brushfield's), blunt inner eye angle, short, broad hands (eg with a single palmar crease), and an incurving 5th digit. Widely spaced first and second toes and a high-arched palate are more obvious later. If you are uncertain about the diagnosis, it is best to ask an expert's help, rather than baffle the mother by taking blood tests 'just in case it's Down's'. *Associated problems:* Duodenal atresia; vsd; patent ductus; ASD (foramen primum defects, p 224); and, later, a low iq and a small stature. Helping the mother accept her (often very lovable) child may be aided by introducing her to a friendly mother of a baby with Down syndrome. *Prenatal diagnosis:* p 166–8.

*Chromosome 21 contains only 225 genes: most of its DNA is apparently meaningless.

Ways of looking at genetic diseases such as Down syndrome

The swot's approach: Learn the clinical manifestations—it might be asked in an exam. In the case of Down syndrome, this person will focus on features such as a simian palmar crease, hypotonia, flat face, upward slanted palpebral fissures and epicanthic folds, speckled irises (Brushfield spots); mental and growth retardation; pelvic dysplasia, cardiac malformations, short, broad hands, hypoplasia of middle phalanx of 5th finger, intestinal atresia, and high arched palate. Associations he or she will learn about include feeding problems, leukaemia, hypothyroidism, and neck joint problems.

The problem with this approach is that it does not make us good doctors because it is not much help to people and their families.

The dangerous young idealist: Down syndrome is preventable. The focus should be on prenatal diagnosis and termination of affected fetuses. Down's cases are unfortunate. These patients are a burden to the State. Their claims on scarce health resources are burdensome. Normal people should take priority.

The problem with this approach is that there are no genetically normal people. This purist has engaged in too much abstract thought: and as Dostoyevsky has taught us: 'too much abstract thought makes men cruel'.

(Crime & Punishment)

The health needs approach: This approach starts with the question: How can I help? Health maintenance for Down's children is more important, not less, compared with the needs of other children—because their families are vulnerable, and many conditions are more likely in those with Down's. Examples are: otitis media, thyroid disease, congenital cataracts, leukaemoid reactions, dental problems, and feeding difficulties.

medline:9930130

Genetic counselling

Goal To provide accurate, up-to-date information on genetic conditions to enable families and patients to make informed decisions.

► Genetic counselling is best done in regional centres to which you should refer affected families. (nearest centre: UK tel.:020 7794 0500) *Don't do blood tests on children lightly which might have long-term consequences, unless some form of treatment is available. The child may never forgive you for labelling him or her.*

In order to receive most benefit from referral:

- The affected person (proband) should come with family (spouse, parents, children, siblings); individuals can of course be seen alone as well.
- The family should be informed that a detailed pedigree (family tree) will be made, and medical details of distant relatives may be asked for.
- Irrational emotions (guilt, blame, anger) are common. Deal with these sensitively, and do not ignore. *Remember:* you do not choose your ancestors, and you cannot control what you pass on to your descendants.
- Warn patients that most tests give no absolute 'yes' or 'no' but merely 'likely' or 'unlikely'. In gene tracking, where a molecular fragment near the gene is followed through successive family members, the degree of certainty of the answer will depend on the distance between the marker and the gene (as crossing-over in meiosis may separate them).
- Accept that some people will not want testing, eg the offspring of a Huntington's chorea sufferer—or a mother of a boy who might have fragile x syndrome, but who understandably does not want her offspring labelled (employment, insurance, and social reasons). Offer a genetic referral to ensure that her decision is fully informed (but remember: 'being fully informed' may itself be deleterious to health and wellbeing, p 169).

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Chromosome nomenclature Autosomes are numbered 1 to 22 roughly in order of size, 1 being the largest. The arms on each side of the centromere are designated 'p' (petite) for the short arm, and 'q' for the long arm. Thus 'the long arm of chromosome 6' is written '6q'.

Chromosomal disorders include Down's (trisomy 21, p 210), Turner's (45,x0, p 759) and Klinefelter's (47, xxy, p 750) syndromes. Many genes are involved when the defect is large enough to be seen microscopically.

Autosomal-dominants Adult polycystic kidney (16p), Huntington's chorea, (4p). A single copy of a defective gene causes damage. Some people inheriting the defective gene are phenotypically normal (=reduced penetrance).

Autosomal recessives Infantile polycystic kidney; cystic fibrosis (7q), β -thalassaemia, sickle-cell (11p). In general, both genes must be defective before damage is seen, so carriers are common. Both parents must be carriers for offspring to be affected, so consanguinity (marrying relatives) increases risk.

X-linked Duchenne muscular dystrophy, Haemophilia A & B, fragile x (p 752). In female (xx) carriers a normal gene on the 2nd x chromosome prevents bad effects manifesting. Males (xy) has no such protection.

NB: being pregnant and unwilling to consider termination does *not* exclude one from undergoing useful genetic counselling.

'Couple screening' A big problem with counselling is the unnecessary alarm caused by false +ve tests. In cystic fibrosis screening (analysis of cells in mouthwash samples) this is reducible by 97% (0.08% vs 3.2%) by screening mother and father together—who need only get alarmed if they *both* turn out to be screen-positive. The trouble with this is false reassurance. Many forget that they will need future tests if they have a different partner, and those who do not are left with some lingering anxieties.

Should genetic counsellors try to influence pregnancy outcome?

Two contrasting principles:

- 1) The parents must decide: counselling must be nondirective.
- 2) Every newborn child has the right to be born healthy, if possible.

Nondirective counselling is something of a mantra among counsellors, partly as a reaction to Nazi eugenic excesses, and partly because of an unwillingness to promulgate a single view of what is right in what can be a very complex area. Public health doctors are questioning this obedience to the nondirective ethic because, from their point of view, it makes attainment of their chief goal more difficult—namely to improve the health and well-being of all residents, including newborns. For example, we should tell pregnant women not to drink much alcohol, public health doctors assert, because this is necessary to prevent the fetal alcohol syndrome. The same goes for other syndromes. When we know what to do we should state clearly and unequivocally what the mother should do. This is our duty to her and her unborn child.

Let us examine the public health doctor's stand point more closely. He wants to improve health. To do this, it is necessary to define health. We have done this elsewhere (p 414) and have argued that health entails more than just soundness of body and mind. It is not clear that an autocratic society in which patients were told what to do would be more healthy than a society of autonomous individuals each addressing the great questions of health and existence from his or her own view point.

Another problem for the public health doctors is that, in the case above, it is not clear whether the directive 'don't drink if you are pregnant' would lead to fewer children with the fetal alcohol syndrome. It might lead to more orphans (a mother knows she is doing wrong, feels guilty, avoids health professionals, and dies of some unforeseen consequence of pregnancy, or from guilt-borne suicide).

medline:10943801

As ever, the way forward is not by abstract thought but by getting to know our patients better. There may be rare occasions when we know our patients well enough to risk 'You are mad not to follow this advice...'. But mostly we cannot be sure that this injunction will work, and it is wiser to explore the patient's world view and their expectations, and then use this knowledge to reframe the benefits of your proposed action in a way that makes sense to the individual concerned.

Measles, rubella, mumps, and erythroviruses

Measles *Cause:* RNA paramyxovirus. *Spread:* droplets. *Incubation:* 7–21 days. Infective from prodrome (catarrh, wretchedness, conjunctivitis, fever) until 5 days after rash starts. Conjunctivae look glassy; then the semilunar fold swells (Meyer's sign). *Koplik spots* are pathognomonic, grain-of-salt-like spots on buccal mucosa. They are often fading as the rash appears (starts behind ears, eg on days 3–5, then spreads down the body, becoming confluent).

Complications: Febrile fits, otitis media, meningitis, *D&V*, keratoconjunctivitis; immunosuppression; subacute sclerosing panencephalitis, if immunosuppressed, giant cell pneumonitis. Intrauterine infection may cause malformations. The worst complication is the encephalitis (headache, lassitude, fits, coma)—up to 15% may die; 25% develop fits, deafness, or ↓ cognition.

Treatment: Isolate—in hospital, if the patient is ill or immunocompromized or malnourished, or has pneumonitis, CNS signs, or dehydration, then:

- Ensure adequate nutrition (catabolism is very high). Continue breast feeding, even during diarrhoea. Pass a nasogastric feeding tube if intake is poor.
- Frequent mouth washes with water + 1% gentian violet to mouth lesions.
- In the developing world, need for vit. A rises; consider 2 doses, + 1 more at 6 weeks later (p 512). *CI:* pregnancy; known not to be deficient.
- Treat 2° bacterial infection, antibiotics such as amoxicillin for otitis media and pneumonia. Prophylactic antibiotics have no clear rôle. medline:9040311

Immunization: p 208. *Prognosis:* Mostly excellent in rich countries—but in poor areas death rate is ~10% (0.9 million/yr, mostly in Africa). B. Beilix Lancet 2002 1564

Rubella (=german measles, german being *germane* ie closely akin to) *Cause:* RNA virus. *Incubation:* 14–21 days. *Infectivity:* 5 days before & 5 days after the rash starts. *Signs:* Usually mild; macular rash; suboccipital lymphadenopathy.

Treatment: Symptomatic. *Immunization:* Live virus, p 208. medline:11348695

Complications: Small joint arthritis. Malformations *in utero* (p 98). Infection during the first 4 weeks: eye abnormalities in 70%; during weeks 4–8: cardiac abnormalities in 40%; weeks 8–12: deafness in 30%.

Mumps (RNA paramyxovirus) *Spread:* Droplets/saliva. *Incubation:* 14–21d.

Immunity: Lifelong, once infected. *Infectivity:* 7 days before and 9 days after parotid swelling starts. *Symptoms:* Prodromal malaise; fever; painful parotid swelling, becoming bilateral in 70% (other causes: Sjögren's; leukaemia; dengue; herpesvirus; EBV; HIV; sarcoidosis; pneumococci; haemophilus; staphs; anaphylaxis; blowing glass or trumpets; drugs). medline:11073786 & 9175423

Complications: Orchitis (± infertility), arthritis, meningitis, pancreatitis, myocarditis, deafness, myelitis. Complete recovery is the rule. medline:10983392

Treatment: Rest. *Immunization:* MMR vaccine, p 209.

Erythrovirus (= parvovirus) Type B19 is the cause of 'slapped cheek' syndrome, also known as 'fifth disease' and erythema infectiosum. This is usually a mild, acute infection of children, characterized by malar erythema (a raised, fiery flush on the cheeks) and a rash mainly on the limbs. Constitutional upset is mild. Arthralgia is commoner in adults—who may present with a glandular fever-like syndrome and a false +ve Paul–Bunnell test. medline:8179710

Spread is rapid in closed communities. It is also the cause of aplastic crises in sickle-cell disease. It is uncommon in pregnancy but risks fetal death in ~10% (especially mid-trimester), medline:9255087 and is a cause of hydrops fetalis by inhibition of multiplication and lysis of erythroid progenitor cells (monitor AFP p 166, for several weeks; if it rises abnormally, do ultrasound). Fetal/neonatal problems do occur: hydrops (occurs in ~3%; treat by intrauterine transfusion if severe), growth retardation, meconium peritonitis, myocarditis/ infarction, glomerulonephritis, placentomegaly, hepatomegaly, oedema, rashes, anaemia,

platelets/wcc↓. Respiratory insufficiency and deaths have been reported, but are rare. ~1:10 of those affected before 20 weeks will miscarry; in the remainder, the overall risk of congenital abnormality is ~1%. medline:9501782

Hand, foot & mouth disease The child is mildly unwell; develops vesicles on palms, soles, and mouth. They may cause discomfort until they heal, without crusting. *Incubation:* 5–7 days. *Treatment* is symptomatic. *Cause:* Coxsackievirus A16 or enterovirus 71 (suspect in outbreaks with herpangina, meningitis, flaccid paralysis ± pulmonary oedema). Herpangina entails fever + sore throat + vesicles or macerated ulcers (on palate or uvula, which heal over 2 days) ± abdominal pain and nausea. www.txt.co.uk/full/cfm?1041

Note Measles, rubella, and mumps are notifiable diseases in the UK. MMR vaccine: see p 208–9. **Herpes infections** See *OHCM* p 570.

Roseola infantum This is a common, mild, self-limiting illness in infants (usually <1yr old), causing fever and then a maculopapular rash on subsidence of fever at the end of the 4th febrile day. Uvulo-palatoglossal junctional (UPJ) ulcers may be a useful early sign. medline:10942088 & 10972002

Cause: Herpes virus 6 (HHV-6), a double-stranded DNA virus. It is morphogenetically related to other herpes viruses (HSV 1 & 2, varicella zoster virus, EBV & CMV). Interaction between host immunities and other viruses may lead to cellular immunodeficiency and fatal illness. *Synonyms:* exanthem subitum, fourth disease, three-day fever. It is neurotropic (a rare cause of encephalitis/focal gliosis on MRI—maybe accounting for why the not uncommon roseola ‘febrile’ seizures tend to occur after the fever). medline:10891638 & 10429479

Other causes of rashes in children See also *skin diseases* section (p 574).

- A transient maculopapular rash is a feature of many trivial viral illnesses (but a few macules may be a sign of early meningococcaemia).
- Purpuric rashes: meningococcaemia (p 258); Henoch–Schönlein purpura (p 275); idiopathic thrombocytopenic purpura (check FBC).
- Drug eruptions: maculopapular rashes in response to penicillins and to phenytoin are particularly common.
- Scabies (p 600); insect bites.
- Eczema (p 588); urticaria (p 578); psoriasis—guttate psoriasis may follow a respiratory tract infection in children (p 586); pityriasis rosea (p 594).
- Still’s disease: transient maculopapular rash, fever, and polyarthritis (p 758).

Managing fever in viral illnesses

Paracetamol syrup (120mg/5mL) is for high fever or severe discomfort. Dose (6hrly po): <3 months old: 5–10mg/kg (a loading dose of ≤40mg/kg PR may be needed); 3 months–1yr: 60–120mg; 1–5yrs: 120–250mg; 6–12yrs 250–500mg. 60mg suppositories are available. Tepid (not cold) sponging augments paracetamol. Also unwrap and rehydrate (this avoids vasoconstriction). Usually antipyretics are not required, and there is evidence of increased mortality if used in severe sepsis. medline--9596591 & 7845113

Varicella (herpes) zoster virus (vzv)

Chickenpox is a primary infection with varicella-zoster virus. Shingles (*OHCM* p 570) is a reactivation of dormant virus in the posterior root ganglion.

Chickenpox *Presentation:* Crops of vesicles of different ages on the skin, typically starting on back. *Incubation:* 11–21 days. *Infectivity:* 4 days before the rash, until all lesions have scabbed (1 week). Spread is by droplets. It can be caught from someone with shingles. It is one of the most infectious diseases known. 95% of adults have been infected; immunity is life-long.

Tests: Fluorescent antibody tests and Tzanck smears are rarely needed.

Differential: Hand, foot, and mouth disease; insect bites; scabies; rickettsia.

Course: Starts as fever. Rash starts 2 days later, often starting on the back: macule→papule→vesicle with a red-surround→ulcers (eg oral, vaginal)→crusting. 2–4 crops of lesions occur during the illness. Lesions cluster round areas of pressure or hyperaemia.

Dangerous contexts: Associated immunosuppression; cystic fibrosis; severe eczema; neonates. *Treatment:*

Keeping cool may reduce lesion numbers. Trim nails to lessen damage from scratching. Daily antiseptic for spots may be tried (chlorhexidine).

Flucloxacillin 125–250mg/6h po if bacterial superinfection—treat for septicaemia if deteriorating. Antivaricella-zoster immunoglobulin (12.5u/kg im max 625u) + aciclovir if immunosuppressed or on steroids and within 10 days of exposure (give as soon as possible, see *BNF*).

Aciclovir is licensed as a 7-day course in chickenpox; begin within 24h of the rash. *Aciclovir child dose:* (20mg/kg/6h po), eg <2yrs: 200mg/6h po; 2–5yrs: 400mg/6h po. ≥6yrs: 800mg/6h. Tablets are 200, 400, or 800mg; suspension: 200mg/5mL or 400mg/5mL. In renal failure, ↓ dose. There is no clear evidence on aciclovir ↓ complications if immunocompetent, but it may help severe symptoms, eg in older patients, or 2nd or 3rd family contacts. If used, use at the first sign of infection.

Complications: If spots are blackish (purpura fulminans) or are coalescing and bluish (necrotizing fasciitis), get urgent help on ITU. *medline:10799624* Be alert to pneumonia, meningitis, myelitis, CNS thrombi, DIC, LFTs↑, Guillain–Barré, Henoch–Schönlein, nephritis, pancreatitis, myositis, myocarditis, orchitis, and ataxia.

If susceptible, live attenuated vaccine pre-cytotoxics/steroids may be wise.

Shingles *Treatment:* Oral analgesia. Ophthalmic shingles: p 484. Aciclovir may reduce progression of zoster in the immunocompromised (may be rampant, with pneumonitis, hepatitis, and meningoencephalitis).

Aciclovir ivi dose: 10mg/kg/8h (over 1h), with concentration <10mg/mL.

Varicella in pregnancy Pneumonitis and encephalitis are no commoner in pregnancy, despite pregnancy being an immunocompromised state (1 in 400 and 1 in 1000, respectively). Infection within the first 20 weeks (esp. 13–20 weeks) causes congenital varicella in 2%.

medline:11190597

Signs of vzv fetopathy are variable, eg cerebral cortical atrophy and cerebellar hypoplasia, manifested by microcephaly, convulsions and IQ↓; limb hypoplasia; rudimentary digits ± pigmented scars. Maternal shingles is not a cause. If the mother is affected from 1 week before to 4 weeks after birth, babies may suffer severe chickenpox. Give the baby zoster immune globulin 250mg im at birth; if affected, isolate from other babies, and give aciclovir.

Infection is preventable by pre-pregnancy vaccination with live varicella vaccine, but testing for antibodies pre-conceptually is expensive, and cost-effectiveness depends on local rates of seronegativity. ~80% of those who cannot recall any previous chickenpox are, in fact, immune.

medline:8762292

Varicella zoster globulin prevents infection in 50% of susceptible contacts, eg 1000mg im. Infection in pregnancy merits aciclovir (it's probably ok for the fetus). Chickenpox at birth is a problem. Barrier nursing mothers causes distress and is of unproven value; infant's mortality may be 20%.

medline:8819427

HIV infection Prenatal HIV: p 98, HIV in labour: p 155, Adult HIV: OHCM p 580-7.

Consider HIV in any child or infant with recurrent or odd infections, or unexplained organomegaly or lymphadenopathy. Most of the millions of HIV+ve children live in Africa, where transmission rates from mother to baby (vertical transmission) are twice as high as in the West. This may be due to concurrent infections, virulence differences, and frequencies of breast feeding (if an HIV +ve mother breast feeds, this ↑risk of vertical transmission by 14%). Risks and benefits of breast feeding need explaining to mothers. medline:11089813

If reliable water is on tap, with sufficient formula feed, then this may be preferable. Infection can occur from the 1st trimester; ~50% of infections occur around the time of delivery, and are more likely if mothers have symptomatic HIV infection or a high viral load. In a cohort of 330 HIV +ve mothers, those with the p24 antigen had a 3-fold risk of transmission compared with those who were p24 -ve. For the rôle of Caesarean section and zidovudine (AZT) in preventing vertical transmission, see p 98. PCP (below) and CMV are potentially fatal in infants whose mother's HIV status wasn't recognized in pregnancy. Breast feeding ↑risk of combined PCP/CMV infection. medline:11273213

Diagnosing HIV-1 Don't use standard tests (placentally transferred IgG lasts for ≤18 months). Virus culture and PCR (polymerase chain reaction) are reliable, and more sensitive than finding p23 antigen in blood. HIV-specific IgA doesn't cross the placenta, but only occurs in 50% of infected infants <6 months old. Discuss with the lab. ►Aim to diagnose 95% of infected infants before the age of 1 month. Monitoring CD4 counts (OHCM p 580) is important in staging HIV.

Presentation Failure to thrive, feeding problems, fever, diarrhoea, repeated admissions for gastro-enteritis, hepatosplenomegaly, PUO, parotitis, lymphadenopathy, chest infections slow to clear, pneumonia, clubbing, *pneumocystis carinii*, toxoplasmosis, tuberculosis, cryptococcosis, histoplasmosis, CMV infection, lymphoid interstitial pneumonitis, and heart defects.

Lymphoid interstitial pneumonitis (LIP): Signs: tachypnoea, hypoxia, clubbing, diffuse reticulonodular infiltrates on CXR, bilateral hilar lymphadenopathy. It is not an AIDS-defining illness, and it is 5-fold less serious than pneumocystosis.

Prognosis Mean age at death: 9.4yrs. By 3yrs old, up to half with early-onset opportunistic infection have died, vs 3% of those with no such infection. Children with slow progression of HIV have persistent neutralizing antibodies. Transplacental passage of maternal neutralizing antibody may also have a rôle.

Guidelines for preventing pneumocystosis (PCP) with co-trimoxazole

Age / HIV status	PCP prophylaxis	Do CD4 cell counts at:
0-5wks, HIV exposed	No	1 month
5wks-4 months, HIV exposed	No	3 months
4-12 months: -HIV infected	Yes	6, 9, and 12 months
-Status unknown	Yes	6, 9, and 12 months
-No HIV infection	No	No monitoring needed
1-5 yrs, HIV infected	Yes, if CD4 < 500 per µL or CD4 percentage < 15%	At least 3-4 monthly
6-12yrs, HIV infected	Yes, if CD4 < 200 per µL or CD4 percentage < 15%	At least every 3-4 months

Ensure a full course of vaccines (include *Haemophilus* & *Pneumococcus*; avoid live vaccines if very immunocompromised; avoid BCG in children with symptomatic HIV in areas where TB prevalence is low). medline:11289006 & 10580195

Treatment of HIV and of opportunistic infections Refer to a specialized HIV centre. Regimens are constantly being refined, and new drugs are under trial: see *Anti-HIV agents*: OHCM p 586: there is some evidence that children respond in a similar way to adults (eg didanosine is better than AZT).

Some GI malformations

Meckel's diverticulum See p 754.

Tracheo-oesophageal fistula (\pm oesophageal atresia) *Prenatal signs:* Polyhydramnios; small stomach. *Postnatal:* Cough, airway obstruction, \uparrow secretions, 'blowing bubbles', distended abdomen, cyanosis, recurrent pneumonias. *Diagnosis:* Inability to pass a catheter into the stomach; x-rays show it coiled in the oesophagus. Avoid contrast radiology. *Treatment:* Correction of fistulae, using a cervical incision. 2001 medline:11329594

Diaphragmatic hernia *Signs:* Difficult resuscitation at birth; respiratory distress; bowel sounds in one hemithorax (usually left so heart is best heard on the right). Cyanosis augers badly (\therefore lung hypoplasia). Associations: other malformations (neural tube in 50%); trisomy 18; chromosome deletions eg at 15q2, Pierre Robin (p 226). *medline:11288119 Incidence:* 1:2200. *Diagnosis:* Prenatal: ultrasound; postnatal: CXR. *Treatment:* •*Prenatal:* Fetal surgery is not usually practical or available (tracheal obstruction may be tried: it encourages lung growth, so pushing out other viscera)—but premature birth may be caused. •*Postnatal:* Insert a large bore nasogastric tube when diagnosis suspected. Face masks are contraindicated (so intubate and ventilate, if needed, with low pressures at a rate of 100/min and minimum positive end-expiratory pressure). Get surgery in an appropriate centre.

Hirschsprung's disease Congenital absence of ganglia in a segment of colon (or in the rare 'long-segment' disease, all the way up to the stomach) leading to infrequent, narrow stools, GI obstruction, and megacolon. Faeces may be felt *per abdomen*, and PR exam may disclose only a few pellet-like faeces. $\sigma/\phi \approx 3:1$. *Complications:* GI perforation, bleeding, ulcers, enterocolitis (may be life-threatening). *Tests:* barium enema or sigmoidoscopy/biopsy of the aganglionic section, staining for acetylcholinesterase-positive nerve excess). Excision of the aganglionic segment is usually needed. This does not always lead to immediate recovery.

Hydroceles in infancy A processus vaginalis patent at birth, and allowing only fluid from the peritoneal cavity to pass down it, generally closes during the first year of life—so no action is usually needed.⁵ If the fluid-filled sac is adjacent to the spermatic cord, it is called an encysted hydrocele or a spermatic cord cyst. If the proximal opening of the processus vaginalis is wide, a true inguinal hernia is formed, and action is always required. *medline:11125413*

Inguinal hernias These are due to a patent processus vaginalis (the passage which ushers the descending testicle into the scrotum). They present as a bulge lateral to the pubic tubercle. They may be intermittent, appearing during crying. In 25% there are bilateral hernias. The aim is to repair these promptly as there is risk of incarceration. Note: there is often an associated hydrocele, and this may be difficult to distinguish from an incarcerated hernia—exploration is required if there is doubt.

Imperforate anus Most girls have a posterior fourchette fistula, boys having a posterior urethral fistula (may pass meconium in urine). Absence of perineal fistula in boys indicates communication with the urethra (\therefore colostomy may be required). Do GU imaging to show commonly associated GU abnormalities. Posterior sagittal anorectoplasty is possible. medline:9563025

Mid-gut malrotations ►Bilious neonatal vomiting merits immediate surgical referral (pass NGT). *medline:10821158* Absent attachment of the small intestine mesentery can cause mid-gut volvulus or obstruction of the third part of the duodenum by fibrotic bands. Presentation may be late; passage of blood per rectum heralds mid-gut necrosis—and is an indication for emergency surgical decompression. See *OHCM* p 476 for acute gastric volvulus.

Anterior abdominal wall defects—Gastroschisis: A paraumbilical defect with evisceration (extrusion of viscera) of abdominal contents. *Incidence:* ~1.6/10,000; rising, and possibly caused by a vascular event associated with cocaine use. The typical pregnancy is in a young, socially disadvantaged primip. There is usually *no* other defect, and corrective surgery has a good outcome in 90% (so deliver where there are good paediatric surgical facilities, if diagnosed prenatally—aim to diagnose at ultrasound). medline~8876090 &: 8876081

Exomphalos: This is a ventral defect with herniation of abdominal contents into the base of the umbilical cord. There is a covering with peritoneum. *Incidence:* ~4.3/10,000. Other abnormalities are often present. medline:11036795

Genitourinary diseases

Preauricular tags are markers of GU problems; consider GU US scan. medline:10799625

Undescended testis—cryptorchidism (2–3% of neonates, 15–30% of prems; bilateral in 25% of these). On cold days retractile testes may hide in the inguinal pouch, eluding all but the most careful examination (eg while squatting, or with legs crossed, or in a warm bath they may be 'milked' down into position). These retractile testes need no surgery. If truly undescended it will lie along the path of descent from the abdominal cavity. Early (eg at 1 year) fixing within the scrotum (orchidopexy) may prevent infertility and aids later diagnosis of neoplasia (untreated, risk is \uparrow >5-fold). Intranasal gonadotrophin-releasing hormone is unreliable. **NB:** biopsy may cause later malignancy. medline-9169396

Urethral valves present with oligohydramnios or absent or feeble voiding (\pm uraemia and a palpable bladder). *Micturating cystogram:* posterior urethral dilatation. Laser resection is possible. *Antenatal diagnosis:* uss (GU dilatation).

Hypospadias (Narrow meatus on ventral penis). Avoid circumcision: use foreskin for preschool repair; attend to aesthetic considerations. medline:11084694

Epispadias (Meatus on dorsum of penis). May occur with bladder extrophy.

Some congenital and genetic disorders *Horseshoe kidney (crossed-fused kidneys):* Symptoms: Silent or obstructive uropathy \pm renal infections. uss diagnosis: kidneys 'too medial'; lower pole 'too long'; anterior-rotated pelvis; poorly defined inferior border; isthmus often invisible. medline:10625187 *Infantile polycystic kidneys* may cause obstructed labour and respiratory difficulties, with later uraemia and $\text{BP}\uparrow$. Radiology: collecting tubule cysts (<5mm across). The histology of the liver is always abnormal. Survivors risk UTIs and portal hypertension with haematemesis. *Inheritance:* recessive (dominant in adult polycystic disease, *OHCM* p 276). medline:7554350

Ectopic kidney: May be seen on us scan (eg pelvic mass) or renal scintigraphy. Associations: anorectal abnormalities, UTIs; calculi. medline:11105630

Renal agenesis causes oligohydramnios, Potter's facies (p 120) + death if bilateral. **VACTERL** association (vertebral, anal, cardiac, tracheoesophageal, renal and limb anomalies). *Diagnosis:* prenatal us scan. medline 11281494

Patent urachus: Urine leaks from the umbilicus. *Image:* excretory urogram.

Bladder extrophy: Pubic separation with bladder exposure. medline:11216679

Double ureter: Associations: ureterocele, UTI, pyelonephritis; may be symptomless.

Renal tubular defects: (eg renal glycosuria, cystinuria, or diabetes insipidus). In *renal tubular acidosis* conservation of fixed base is impaired, causing metabolic acidosis + alkaline urine. *Symptoms:* Failure to thrive; polyuria; polydipsia.

Wilms' nephroblastoma This is the commonest intra-abdominal tumour of childhood (20% of all malignancies). It is an undifferentiated mesodermal tumour of the intermediate cell mass. It may be sporadic, or familial (2%), or associated with Beckwith–Wiedemann syndrome (BWS, p742), aniridia, GU malformations, and retardation (WAGR). One of the Wilms' tumour genes (WT1 on chromosome 11) encodes a protein which is a transcriptional repressor downregulating IGF-II, an insulin-like growth factor. medline.7707816 Median age at presentation: 3.5yrs. 95% are unilateral. Staging:

- | | |
|--------------------------------------|-----------------------|
| I Tumour confined to the kidney | IV Distant metastases |
| II Extrarenal spread, but resectable | V Bilateral disease |
| III Extensive abdominal disease. | |
- www.vh.org/Providers/TeachingFiles/RCVW/030196/WilmsTumour.html

The patient: Features include fever, flank pain, an abdominal mass. Haematuria is not common. Ultrasound: renal pelvis distortion; hydronephrosis.

Management: Avoid biopsy; nephrectomy + vincristine & actinomycin 4 weeks pre-op can cure. Radiotherapy may help. Genetic & biological factors guide risk categorization and help individualize care. 2001 medline 11208843 & 11137681

Ambiguous genitalia (intersexuality)

This is rare: *refer promptly*. ►Distinguish genetic, gonadal, phenotypic (affected by sex hormone secretion etc.), psychological, and social-rôle sexualities.

Male sex differentiation depends on SRY genes (on Y chromosomes) transforming an indifferent gonad into a testis; its products (testosterone & Mullerian inhibiting substance) control fetal sex differentiation.* *medline:9059565*

History & examination Any exposure to progesterone, testosterone, phenytoin, aminoglutethamide? Previous neonatal deaths (adrenogenital syndrome is recessive)? Note phallic size, and position of the urethral orifice. Are the labia fused? Have the gonads descended? Undescended palpable testes are more likely to signify an intersex condition than palpable undescended testes—likewise with severity of hypospadias. *medline:10458421*

Tests Buccal smear (Barr body suggests ♀); WBC mustard stains make Y chromosomes fluoresce; these take <24h vs 5 days for chromosome analysis. If there is a phallus and buccal smear is 'female' diagnosis is either adrenogenital syndrome or maternal androgens (drugs, tumours) or true hermaphroditism, ie ovarian and testicular tissue coexist, eg with a different gonad on each side (46,XY, 46,XX, 45,XO/46,XY or 45,X/47,XXY mosaic). *medline:11327376*

If there is a phallus and buccal smear is ♂, tell mother that the baby is a boy.

Do not rely on appearances whenever babies have: bilateral cryptorchidism (at term), even if a phallus is present; unilateral cryptorchidism with hypospadias; penoscrotal or perineoscrotal hypospadias. These patients need examination by a paediatric endocrinologist to exclude androgen resistance (eg testicular feminization).* Genetic tests are also vital: eg terminal deletion of 10q deletes genes essential for normal male genital development. *medline:9933844*

NB: if the stretched phallus is <25mm long, normal procreation is unlikely. If there is uncertainty due to a short penis, a paediatric endocrinologist may try 3 days' treatment with human chorionic gonadotrophin. If the baby is a boy the penis will grow (possibly even to normal length) after 5 days. *medline:8393754*

Aromatase deficiency CYP19 genes are needed for normal oestrogenization: recessive mutations cause ambiguous genitalia in 46,XX individuals; at puberty there is hypergonadotropic hypogonadism, with no secondary sexual characteristics, except for progressive virilization. Boys have normal male sexual differentiation but are tall and brittle (↓epiphyseal closure ± osteoporosis from oestrogen lack). Oestrogen receptor gene mutations are similar. *medline:11305285*

Congenital adrenal hyperplasia CAH is due to excess secretion of androgenic hormones because of deficiencies of 21-hydroxylase, 11-hydroxylase, or 3-β-hydroxysteroid dehydrogenase. Cortisol is inadequately produced, and the consequent rise in adrenocorticotrophic hormone leads to adrenal hyperplasia, and overproduction of androgenic cortisol precursors. CAH is a leading cause of ♀ pseudohermaphroditism. *Incidence: 1:14000. medline:11242695*

Clinical & biochemical features: Vomiting, dehydration, and ambiguous genitalia. Boys may seem normal at birth, but will exhibit precocious puberty, or may have ambiguous genitalia (↓androgens in 17-hydroxylase deficiency), or incomplete masculinization (hypospadias with cryptorchidism from 3β-hydroxysteroid dehydrogenase deficit). Hyponatraemia, (with a paradoxically high urine Na⁺) and hyperkalaemia are common. ↑Plasma 17-hydroxyprogesterone in 90%; urinary 17-ketosteroids ↑ (not in 17-hydroxylase deficit).

Emergency treatment: Babies may present with an adrenocortical crisis (circulatory collapse) in early life. Other presentations include hyponatraemic seizures in infancy (often misdiagnosed as a febrile convulsion). ►►Urgent treatment is required with 0.9% saline iv (3–5g Na⁺/day), glucose, and hydrocortisone 100mg/2–6h iv (all ages). Give fludrocortisone 0.1mg/day po.

Assigning sex and gender

► Do not shy away from telling patients that you do not know whether their baby is a boy or girl, and that tests must be done (it may be wise to await results before naming the child). This is unsatisfactory, but much better than having to reassign gender. This is why a neonate with ambiguous genitalia is a sexual emergency. Choice of gender must take into account chromosomal and gonadal sex, the hormonal milieu during fetal life, surgical aspects, internal anatomy, fertility issues, psychosexual development, and adult sexual function. NB: ♀ karyotype does not guarantee absence of intra-abdominal testes—so future risks of malignancy have to be assessed too.

medline:11051704

Your job may be to assemble an optimum team: ie a paediatric endocrinologist with psychological expertise, and a laparoscopic surgeon skilled in neonatal cystoscopy and genitography as well as in reconstruction.

Prenatal preparation entails comparing prenatal karyotype with us genital scans to formulate an intersex differential diagnosis—but us scans are unreliable in >50% of ♀ pseudohermaphroditism. medline:10754161 & 10958723

NB: It is common to assign female gender when in doubt, medline:11149399 but some experts favour a gender compatible with the chromosomal sex, if at all possible. This too is a simplification as we don't fully understand determinants of gender rôle (social sex). medline:11039078 & 11242664

Advise against registering the birth until a definite treatment plan is in place. Once registered, legal sex cannot be changed in most countries.

Genetics, biosynthetic pathways, and the possibility of screening for congenital adrenal hyperplasia

The common cause of adrenal hyperplasia is one of ~10 gene defects (6p21.3; the gene is called *CYP21*) that cause deficiency of 21-hydroxylase (which mediates the penultimate step in cortisol biosynthesis). Corticotrophin-induced accumulation of 17OH-progesterone leads to ↑ levels of testosterone (via hepatic conversion). Most affected infants are also salt losers, as 21-hydroxylase is needed for aldosterone biosynthesis, and in boys this is usually the sole early manifestation (excess virilization may be early or delayed to adulthood). Biochemical screening is rarely universal, but some centres use it in boys, aiming for diagnosis before life-threatening adrenal hyperplasia. (Girls are generally detected by finding virilization at neonatal examination providing the baby is not misdiagnosed as a boy—also prenatal diagnosis is possible, and treatable by giving the mother dexamethasone from early in pregnancy.)

One difficulty is the diversity in time of onset and clinical presentation despite identical *CYP21* mutations, making adrenal hyperplasia a continuum of disorders. Treatment is medical and surgical (clitoral reduction and vaginoplasty). Growth and fertility are also impaired. medline:9737273

Androgens cause maleness in 46,XY individuals, provided no mutations in the X-linked androgen receptor occur. Mutations cause more or less complete androgen insensitivity and female phenotype, with normal levels of testosterone and dihydrotestosterone (DHT). If there is partial insensitivity, topical (periscrotal) DHT (Andractim®) has been used to augment maleness; but if *in vitro* functional assays show this is impossible, babies are usually brought up as girls. medline:10543676

Congenital heart disease (CHD)

Incidence 8/1000 births. 2 key questions: *Is the defect compensated?* (unobtrusive); if not, *Is there cyanosis?* **Signs of decompensation:** Poor feeding, dyspnoea, hepatomegaly, engorged neck veins, pulse \uparrow . (or \downarrow , premorbidly) **Physiological types:** • Left to right shunt (ASD; VSD; AVM) • \downarrow Systemic perfusion: weak pulse \pm acidosis (hypoplastic L heart; aortic stenosis; coarctation; any left outflow obstructions) • Pulmonary venous congestion (eg mitral stenosis) • Transposition streaming (of caval return \rightarrow aorta) • \downarrow Pulmonary blood flow (eg Fallot's) • Intracardiac mixing of 'blue' and 'red' blood (eg truncus arteriosus).

Acyanotic causes of uncompensated defects (left to right shunt) Atrial septal defects (ASD), VSDs, aortopulmonary window, patent ductus (PDA). Later pulmonary hypertension may cause shunt reversal and cyanosis (p 746).

Cyanotic causes (R to L shunt via VSD or ASD.) Fallot's (p 746), transposition of the great arteries (TGA); shunt-associated tricuspid or pulmonary stenosis/atresia.

Tests FBC, CXR, P_aO_2 (in air & 100% O_2), ECG, 3D echocardiogram, cardiac catheter.

VSD: (25% of cases) Symptoms: as above. Signs: harsh, loud, pansystolic 'blowing' murmur \pm thrill; \pm a diastolic apical inflow murmur. ECG: left (or combined) ventricular hypertrophy. CXR: pulmonary engorgement. Course: 20% close spontaneously by 9 months (*maladie de Roger*).

ASD: (7% of cases) Symptoms: above. Signs: widely split, fixed S_2 + midsystolic murmur (esp. at 2nd intercostal space at left sternal edge). CXR: cardiomegaly, globular heart (primum defects). ECG: RVH \pm incomplete R bundle branch block.

Patent ductus: Signs: failure to thrive, pneumonias, heart failure, SBE, collapsing pulse, thrill, $S_2\uparrow$, systolic pulmonary area murmur, or continuous 'machinery hum'. CXR: vascular markings \uparrow , big aorta. ECG: LVH. Dexamethasone in preterm labour helps close PDAs. For neonates, indomethacin or ibuprofen 10mg/kg iv + 2 doses of 5mg/kg all separated by 24h also closes PDAs (beware oliguria). medline:10974130

Coarctation: Aortic constriction leads to difficulty in feeling the femoral pulses, \uparrow BP in the arms (may cause epistaxis), and \downarrow in the legs, absent foot pulses, \pm systolic murmur at the left back, heart failure. Think of this whenever there is shock, eg on days 3–10, when ductus closes. CXR: rib notching (late). ECG: LVH.

Transposition of great arteries (TGA): Cyanosis, CCF, \pm systolic murmur. CXR: egg-shaped heart. ECG: RVH. Balloon atrial septostomy allows oxygenated blood to reach the aorta via the right heart. Correction is possible. medline:11219191

Pulmonary stenosis: Pulmonary thrill with systolic murmur. See OHCM p 134.

Treatment • Treat heart failure in babies with nasogastric feeds, sitting upright, O_2 , furosemide eg 1mg/kg/24h slow iv \pm spironolactone; digoxin 10 μ g/kg/24h po is rarely used. • You may be advised to keep the ductus open in duct-dependent cyanotic conditions with prostaglandin E_1 (0.05–0.1 μ g/kg/min iv); intubate and ventilate if needed before transfer to specialists; during transfer be alert to: $T^\circ\downarrow$; $Ca^{2+}\downarrow$; glucose \downarrow ; hypovolaemia. • Prevent SBE (p 194). Open-heart surgery using hypothermia and circulatory arrest is possible at any age, eg for: Fallot's, VSD, TGA, total anomalous pulmonary venous drainage (in which pulmonary veins drain, eg into the portal system, causing CCF). Balloon valvuloplasty \downarrow need for open surgery in pulmonary stenosis (there is more of a problem with restenosis and residual incompetence with aortic valvuloplasty). It is also employed in coarctation. Examples of palliative surgery: pulmonary artery banding to restrict blood flow; systemic to pulmonary shunts to enlarge an underdeveloped pulmonary arterial tree, before inserting a valve-bearing conduit.

Prenatal screening echocardiography Sensitivity: 88%; specificity and +ve predictive value: 100%. Intrauterine cardiac intervention is possible. medline:8826802

Murmurs and heart sounds in children

Benign flow murmurs (eg parasternal low-frequency 'twangs', in early systole) are heard in up to 80% of children, at some time (eg with fever, exercise, or anxiety). *Still's murmur* is an example, and may be abolished by hyperextension of the back, and neck. Lack of other features distinguishes these from malformations: **no** clubbing; **no** cyanosis; **no** thrills; **no** rib recession; **no** clicks; **no** arrhythmias; normal pulses & apex; **no** failure to thrive). When in doubt, do CXR & ECG; get a skilled cross-sectional echocardiogram.

Questions to ask yourself while listening to the 2nd heart sound (S₂)

- Is it a double sound in inspiration, and single in expiration? (Normal)
- Is S₂ split all the time? (ASD—atrial septal defects)
- Is S₂ never split, ie single? Fallot's; pulmonary atresia; severe pulmonary stenosis; common arterial trunk; transposition of the great arteries (the anterior aorta masks sounds from the posterior pulmonary trunk.)
- Is the pulmonary component (2nd part) too loud? (Pulmonary stenosis)

NB: the 2nd heart sound is more useful diagnostically than the first.

Points to note on hearing murmurs If you have an ear for detail! *Timing:*

- *Ejection systolic* (innocent, or semilunar valve or peripheral arterial stenosis).
- *Pansystolic* with no crescendo—decrescendo (VSD, mitral incompetence).
- *Late systolic*, no crescendo—decrescendo (mitral prolapse, *OHCM* p 130).
- *Early diastolic* decrescendo (aortic or pulmonary incompetence).
- *Mid-diastolic* crescendo—decrescendo (↑atrio-ventricular valve flow, eg VSD, ASD; or tricuspid or mitral valve stenosis). An opening snap (*OHCM* p 130) and presystolic accentuation suggest the latter.
- *Continuous murmurs* (PDA, venous hum, or arterio-venous fistula).

Loudness: The 6 grades for systolic murmurs: ▶ Thrills mean pathology.

1 Just audible with a quiet child in a quiet room. 2 Quiet, but easily audible.

3 Loud, but no thrill.

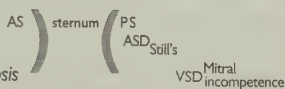
4 Loud with thrill.

5 Audible even if the stethoscope only makes partial contact with skin.

6 Audible without a stethoscope.

Place:

AS = aortic stenosis; PS = pulmonary stenosis



Accentuating/diminishing manoeuvres: (Use our *Oxford Clinical Mentor* to hear the sounds)

- Inspiration augments systemic venous return (the 'negative' pressure draws blood from the abdomen into the thorax), and therefore the murmurs of pulmonary stenosis and tricuspid regurgitation.
- Expiration augments pulmonary venous return and decreases systemic return, and therefore VSD, mitral incompetence, and aortic stenosis too. In (mild) pulmonary stenosis, the ejection click is augmented by expiration.
- Valsalva manoeuvre ↓ systemic venous return and benign flow murmurs, but ↑ murmurs from mitral incompetence and subaortic obstruction.*
- Sitting or standing (compared with lying) ↓ innocent flow murmurs, but ↑ murmurs from subaortic obstruction or from a venous hum (places to listen: right base; below left clavicle; neck—it is abolished by gently pressing the ipsilateral jugular; PDA murmurs are similar, but do not change with posture).

Catheter findings For images, see www.kumc.edu/kumcpeds/cardiology/pedcardiodiagrams.html

Pulmonary stenosis

+ foramen ovale

VSD

ASD

Patent ductus

Fallot's tetrad

RV pressure ↑; pulmonary artery pressure ↓

as above with right atrial pressure ↑ and P_aO₂ ↓

RV pressure ↑; RV O₂ > R atrial O₂

right atrial pressure and oxygenation ↑ compared to ivc

RV pressure ↑; pulmonary artery O₂ > RV O₂

see p 746; RV O₂ & P_aO₂ ↓

Orofacial clefts (cleft lip and palate)

This is the commonest facial malformation. It results from failure of fusion of maxillary and premaxillary processes (during week 5). The defect runs from lip to nostril. It may be bilateral, when there is often a cleft in the palate as well with the premaxillary process displaced anteriorly. Palate clefts may be large or small (eg of uvula alone). *Incidence*: ~1/1000. *Causes*: Genes, benzodiazepines, antiepileptics, rubella. Other malformations are common, eg trisomy 18, 13–15, or Pierre Robin short mandible (causing cyanotic attacks). *Prevention*: Folic acid $\geq 6\text{mg/day}$ periconceptually \pm multivitamins. www.text.co.uk/full/cfm?10

Treatment: A long-term team approach is best (orthodontist, plastic surgeon, oral surgeon, GP, paediatrician & speech therapist). Feeding with special teats may be needed before plastic surgery (usually, repair of the lip is at 3 months and the palate at 1yr; some surgeons do the lip at 1 week old). Repair of unilateral complete or incomplete lesions often gives good cosmesis. Try to refer to expert centres. If bilateral, there is always some residual deformity.

Complications: Otitis media, aspiration pneumonia, post-op palatal fistulae, speech defects (get speech therapy help). Avoid taking to NICU/SCBU (?hinders bonding)

Other malformations in the head and neck

Eyes Anophthalmos: there are no eyes; rare; part of trisomy 13–15.

Ectopia lentis: presents as glaucoma with poor vision. The lens margin is visible; seen in Marfan's, Ehlers–Danlos (p 746), homocystinuria; incidence $< 1/5000$; autosomal-dominant (a-Dom) or recessive (a-R).

Cataract: rubella, Down's, others: recessive or sex-linked.

Coloboma: notched iris with a displaced pupil; incidence: 2/10,000; (a-R).

Microphthalmos: small eyes; 1/1000; due to rubella—or genetic (a-Dom).

Ears Accessory auricles: seen in front of the ear; incidence: 15/1000.

Deformed ears: Treacher–Collins' syndrome (p 758).

Low-set ears: associations—Down syndrome; congenital heart disease.

Nose/throat *Choanal atresia*: Signs: postnatal cyanotic attacks; nasal catheter doesn't go into the pharynx because of nasal malformation. *Incidence*: $\leq 1/5000$.

Surgery: consider a micro-endoscopic nasal approach. 2001 medline:11345154

Congenital laryngeal stridor: (∴ laryngeal webs or laryngomalacia: the larynx is unable to stay open). Signs: shrill inspirations; dyspnoea. *Surgery*: endoscopic.

Laryngeal atresia: First breaths don't expand the lungs, which are hypoplastic.

Look for co-existing anomalies. medline:11043429 *Branchial fistula*: these open at the front of sternomastoid (a remnant of the 2nd or 3rd branchial pouch).

Incidence: $< 1/5000$. Branchial and thyroglossal cysts: *OHCM* p 498.

Skull & spine *Brachycephaly*: Short, broad skull from early closure (cranio-stenosis) of the coronal suture; incidence: $< 1/1000$; Down's-associated or a-Dom. *Cleidocranial dysostosis*: No clavicles (so shoulders meet). Slow

skull ossification, no sinuses, high-arched palate; incidence $< 1/5000$; a-Dom. *Craniofacial dysostosis*: Tower skull, beaked nose, exophthalmos. Δ : spiral CT.

Klippel–Feil syndrome (p 752): fused cervical vertebra (so the neck is short).

CNS Hydrocephalus: incidence 0.3–2/1000. Ante- or neo-neonatal injury, infection, or genes (sex-linked) may cause aqueduct stenosis. Dandy–Walker syndrome (p 744); Arnold–Chiari malformation (*OHCM* p 720).

Microcephaly: Causes: genetic, medline:11262728; intrauterine viruses (eg rubella), hypoxia, x-rays, maternal alcohol. *Incidence*: 1/1000. Recurrence risk: 1/50.

Spina bifida and anencephaly See p 228.

Fetal alcohol syndrome The severity depends on how much alcohol the mother has used in pregnancy. Features: microcephaly, short palpebral fissure, hypoplastic upper lip, absent philtrum, small eyes, IQ↓, cardiac malformations.

Acrocephaly: This term may be applied to tower headed conditions.

Arhinencephaly: Congenital absence of the rhinencephalon.

Arthrogryposis: This term implies contracture of a joint.

Brachycephaly: The head is too short, with a flat occiput (see opposite).

Cebocephaly: A small, flat nose with a single nostril below close-set eyes.

Cephalocele: Intracranial contents protrude. 'Kélé' means hernia in Greek.

Cyclopia: A single eye in the area normally occupied by the root of the nose, which is missing, or present in the form of a proboscis (a tubular appendage) located above the eye. In some, it can be viewed as an extreme form of hypotelorism. It may be part of trisomy 13 (Patau's syndrome).

Craniostenosis = craniosynostosis = premature closure of skull sutures.

Dolicephalic: The head is elongated, eg as in Marfan's, or El-Greco portraits.

Dystopia canthorum: Intercanthal distance is increased, but not the interpupillary or (bony) interorbital distances.

Holoprosencephaly: (a whole, ie single-sphered, brain) Hypotelorism with cleft palate \pm premaxillary agenesis \pm cyclopia \pm cebocephaly (see above)—follows failure of the lateral ventricles to separate (defective cleavage of the prosencephalon), eg with fusion of the basal ganglia. 3 types: lobar (often fatal in infancy), semilobar, and lobar, reflecting degree of the cleavage abnormality. Deletions of chromosomes 7q and 3p or trisomy 13, 18, and 21 may be causal (sometimes). There may be mental retardation, epilepsy, quadriplegia, neuroendocrine anomalies, and poor growth.

Lissencephaly: Smooth cortex with no convolutions (agyria), with 4 layers instead of the usual 6. Signs: failure to thrive, microcephaly, developmental delay, seizures, optic nerve hypoplasia, and microphthalmia. In some, it is associated with the Miller–Dieker syndrome (prominent forehead, bitemporal hollowing, anteverted nostrils, prominent upper lip, and micrognathia). The gene LIS-1 gene at 17p13.3 may be deleted. www.isabel.org.uk

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Micrognathia: The mandible is too small.

Metopic suture: This is the same as the frontal suture.

Neurocranium: That part of the skull holding the brain.

Obelion: The point on the sagittal suture crossed by a line joining the parietal foramina.

Oxycephalic = Turricephaly = acrocephaly = the top of the head is pointed.

Plagiocephaly: If fully expressed, there is synostosis affecting a coronal (and occasionally lambdoidal) sutures (\pm palpable bony ridge) with a flat forehead and elevation of the orbit on the affected side. Minor (unfused) plagiocephalic asymmetry is common in early infancy, improves with time, and is of no significance. Associations: scoliosis and pelvic obliquity.

Rachischisis: Congenital fissure of the spinal column.

Inciput: The anterior, upper part of the head.

Viscerocranium: Facial skeleton.

Vormian bones: Supernumerary bones in the sutures of the skull.

Neural tube defects

Myelodysplasia: Any neuroectoderm defect, eg of the cord, either multiple anterior horns, several central canals, failure of cord fusion so that there is a flat neural plaque, not a fused tube (*myelocele*), or a double neural tube (*diplomyelia*), or herniation through a bony defect as a *meningocele* (contains dura & arachnoid) or *meningomyelocele* (the cord is involved too). *Spina bifida* implies an incomplete vertebral arch (*spina bifida occulta* if covered by skin.) *Anencephaly* implies absent skull vault and cerebral cortex. *Encephalocele* means that part of the brain protrudes through the skull.

UK prevalence 5.6/10,000 (half of what it was in 1990). medline:10791652

Risk increases with young primips, lower social class—and homozygosity for a point mutation (C677→T; prevalence ≈ 10%, interfering with folate metabolism). **Recurrence risk** rises 10-fold if one pregnancy is affected, 20-fold if 2, 40-fold if 3 pregnancies affected; and 30-fold if a parent is affected.

Neurological deficit is very variable, depending on the level of the lesion, and the degree to which the lower cord functions independently from the upper cord. The defect may progress after birth (and after subsequent operations); subsequent hydrocephalus gradually worsens mental performance. A child who learns to walk during his 2nd year may outgrow his ability to support himself during the next years (weight increases as the cube of surface area, power only as its square). Those with lumbosacral myelomeningoceles usually learn to walk with calipers by the age of 3, but ≤20% with higher lesions ever walk. An unstable condition exists when there is paralysis below L3, as unopposed hip flexors and adductors are likely to dislocate the hips. Only 5–13% retain their ability to walk. medline:10454227

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Postnatal surgery Firm guidelines on whom to treat often prove too simple to apply to the individual infant. The final outcome of early closure of the defect depends on the state of the kidneys after multiple UTIs, and the extent of delayed hydrocephalus (requiring ventriculoperitoneal CSF shunts). Early post-operative mortality may account for ~25% of deaths. Many operations may be needed for spinal deformity (often severe and very hard to treat).

Intrauterine diagnosis A maternal serum α-fetoprotein >90u/mL at 18 weeks detects ~80% of open spina bifidas, and 90% of anencephalics, but also 3% of normal singleton fetuses, twins, and some with exomphalos, congenital nephrosis, urethral valves, Turner's syndrome, trisomy 13, and oligohydramnios. Amniocentesis and skilled ultrasound ↑pick-up rates further.

Intrauterine surgery (eg at 23 weeks' gestation) This is very controversial.

Hurdles for the developing child

- Urinary and faecal incontinence. Penile appliances, urinary diversions or intermittent self-catheterization (for girls) save laundry and bed sores.
- The mother who 'does it all' can prevent maturity developing.
- Immobility. Mobility allowances are small and of little help.
- Social and sexual isolation, if a special school is needed. medline:782378

Prevention In mothers who have already had an affected baby, there is good evidence that folic acid (5mg/day) given from before conception (as the neural tube is formed by 28 days, before pregnancy may even be recognized) reduces the risk of recurrence of neural tube defects by 72%. If no previous neural tube defects, 0.4mg of folic acid is recommended in the months before conception and for 13 weeks after. See p 94. medline:9400502

Many mothers don't take folate pre-conception, hence the call for folate-fortification of bread etc. This is adopted in the USA, but is not ideal, as pernicious anaemia may be masked. medline:9950465 & 10609896 & 10489953

►► Neonatal life support (NLS) after delivery

Most neonates are perfectly healthy, and the best plan is to return these babies to the mother without interference, to augment bonding. A paediatrician or nurse trained in advanced neonatal resuscitation should attend the following births: emergency Caesareans, breeches, twins, forceps deliveries for fetal distress, intrapartum bleeding, prematurity, hydrops fetalis, and eclampsia.

Before birth Check the equipment. Get a warm blanket. Heat the crib.

At birth If pulse <100, poor colour or respiratory effort, set a clock in motion, and see opposite. Be alert to:

- Hypothermia (use heat lamp).
- Hypoglycaemia—give dextrose 25%, 2mL/kg iv.
- Opiate toxicity—naloxone 200µg IM (100µg if <1.5kg).
- Anaemia (heavy fetal blood loss?)—give 20mL/kg 4.5% albumen + blood.
- Is there lung disease or congenital cyanotic heart disease (p 224)? Transfer to NICU/SCBU for monitoring.
- Suck out the oropharynx only if meconium aspiration suspected.

Endotracheal intubation is a key skill: use 3.5mm uncuffed, unshouldered tubes on term infants; 3 if 2.5–1.25kg (2.5 if smaller). Learn from experts. Have larger and smaller sizes to hand. Practice on models.

Prognosis Mortality for *prems* is 315/1000 if 5-min Apgar score = 0–3, vs 5/1000 if score is ≥7. Corresponding figures *at term* are 244/1000 & 0.2/1000. If a term infant with Apgar score ≤3 has a low arterial pH (≤7), risk of neonatal death increases 8-fold. www.nejm.org/content/2001/0344/0007/0467.asp Survival in those needing CPR (cardiac resus) is 63% for infants of 0.5–1.5kg, compared with 88% in these weight groups if CPR is not needed. Severe intraventricular haemorrhage is seen in 15% of those who need CPR, vs 5% in those who don't. www.txt.co.uk/sum.cfm?1035

Dry the baby; remove any wet cloths and cover.

↓
Initial assessment; set a clock in motion; assess **colour, tone, breathing** and **pulse**.
If not breathing. . .

↓
Control the airway (head in the neutral position)

↓
Support breathing: 5 inflation breaths, each 2–3sec long. Confirm response: visible chest movements or increased heart rate.

↓
If no response, check **head position** and try a **jaw thrust** and then 5 inflation breaths. Confirm response: visible chest movements or increased heart rate.

↓
If still no response, get a 2nd person to help with airway control and repeat inflation breaths.

- Direct vision of oro-pharynx
- Repeat 5 inflation breaths
- Insert an oro-pharyngeal airway
- Repeat inflation breaths

Consider intubation. Confirm response: visible chest movements or increased heart rate.

↓
When chest is moving, continue with ventilation breaths if no spontaneously breathing.

↓
Check the heart rate; if undetectable or <60 (and not rising)

start chest compressions. Do 3 chest compressions to 1 breath, for 30sec

↓
Reassess pulse: if improving, stop chest compressions.
If not breathing, continue ventilation.
If heart rate still slow, continue ventilation and chest compressions.

↓
Consider **iv access** and **drugs**, eg adrenaline (epinephrine): 10µg/kg iv or 20µg/kg via endotracheal tube.

↓
► At all stages...ask: **do I need help?**

Apgar	Pulse	Respiratory effort	Muscle tone	Colour	On suction
2	>100	strong cry	active	pink	coughs well
1	<100	slow, irregular	limb flexion	blue limbs	depressed cor
0	0	nil	absent	all blue or white	no response

Examination of the neonate*

The aim is to screen for abnormality, and to see if the mother has any questions or difficulties with her baby. The following is a recommended routine before the baby leaves hospital—or during the first week of life for home deliveries. Before the examination find out if the birth weight was normal. Was the birth and pregnancy normal? Is mother Rhesus –ve? Find a quiet, warm, well-lit room. Enlist the mother's help. Explain your aims. Does she look angry or depressed? Listen if she talks. Examine systematically, eg from head to toes.

The head Circumference (30–38cm, p 294), shape (odd shapes from a difficult labour soon resolve), fontanelles (tense if crying or intracranial pressure↑; sunken if dehydrated). **Eyes:** Red reflex (absent in retinoblastoma); corneal opacities; conjunctivitis. **Ears:** Shape; position. Are they low set (ie below eyes)? The tip of the nose, when pressed, is an indicator of jaundice in European children. Breathing out of the nose (shut the mouth) tests for choanal atresia. Ensure that oto-acoustic screening is done (p 540). Are follow-up brainstem evoked responses needed? 2001 medline:11283492

The complexion: Cyanosed, pale, jaundiced, or ruddy (polycythaemia)? **Mouth:** Look inside. Insert a finger—is the palate intact? Is suck good?

Arms & hands Single palmar creases (normal or Down's). Waiter's (porter's) tip sign of Erb's palsy of C5 & 6 trunks. Number of fingers. Clinodactyly.

The thorax Watch respirations; note grunting or intercostal recession (respiratory distress). Palpate the precordium and apex beat. Listen to the heart and lungs. Inspect the vertebral column for neural tube defects.

The abdomen Expect to feel the liver and spleen. Are there any other masses? Next inspect the umbilicus. Is it healthy? Surrounding flare suggests sepsis. Next, lift the skin to assess skin turgor. Inspect the genitalia and anus. Are the orifices patent? 93% of neonates pass urine within the first 24h. Is the urinary meatus misplaced (hypospadias), and are both testes descended? The neonatal clitoris often looks rather large. Bleeding PV may be a normal variant following maternal oestrogen withdrawal.

The lower limbs Test for congenital dislocation of the hip (p 632). Avoid repeated tests as it hurts—and may induce dislocation. Can you feel the femoral pulses (to exclude coarctation)? Note talipes (p 632). Are the toes: too many, too few, or too blue?

Buttocks/sacrum Are there 'mongolian spots'? These are blue and harmless. Tufts of hair ± dimples: is there spina bifida occulta? Any pilonidal sinus?

CNS Posture and handling the baby. Now intuition can be most helpful in deciding whether the baby is ill or well. Is he jittery (hypoxia/ischaemia, encephalopathy, hypoglycaemia, infection, hypocalcaemia)? There should be some degree of control of the head. Do the limbs move normally, and is the tone floppy or spastic? Is the response absent on one side (hemiplegia)? The Moro reflex rarely adds important information (and is uncomfortable for the baby). It is performed by sitting the baby at 45°, supporting the head. On momentarily removing the support the arms will abduct, the hands open and then the arms adduct. Stroke the palm to elicit the grasp reflex. Is the baby post-mature, light-for-dates or premature (p 180)?

► Discuss any abnormality with the parents after liaising with a senior doctor.

*The neonatal period is the 1st 28 days of life; if prem, 44 completed weeks of the infant's conceptional age (= the chronological age plus gestational age at birth). [@11358751]

Neonatal intensive care

Neonatal intensive care is a technological development of the basic creed of first aid—ABC; A for airway, B for breathing, and C for circulation. Success depends on mastering this trinity, and designing the best *milieu intérieur* to encourage healing and growth without developing neurodevelopmental sequelae, or spasticity. As more physiological functions are taken over by machines, monitoring the state of the *milieu intérieur* becomes ever more vital. Monitor temperature, pulse, blood pressure (intra-arterial if critical), respirations (continuous read-out device), blood gases—transcutaneous oximetry ($S_{A}O_2$, *OHCM* p 154) and CO_2 monitoring, or via intra-arterial electrode, U&E, bilirubin, FBC, daily weight, weekly head circumference.

The patient is usually a premature baby. His mortal enemies are cold, hypoglycaemia (p 238), respiratory distress syndrome (p 240), infection (p 238), intraventricular haemorrhage, apnoea, and necrotizing enterocolitis (p 244).

Cold With their small volume and relatively large surface area, this is a major problem for the premature and light-for-dates baby. The problem is circumvented by using incubators which allow temperature (as well as humidity and F_iO_2) to be controlled, and also afford some protection against infection. F_iO_2 is the fraction of O_2 in inspired air, ie $0.6=60\% O_2$.

Apnoeic attacks *Prevalence:* 25% of neonates <2.5kg; 90% if <1kg. *Causes:* Respiratory centre immaturity; aspiration; heart failure; infection; $P_aO_2 \downarrow$; glucose \downarrow ; $Ca^{2+} \downarrow$; seizures; PDA; $T^\circ \uparrow$ or \downarrow ; exhaustion; airway obstruction. If stimulating the baby doesn't restore breathing, suck out the pharynx and use bag and mask ventilation. Avoid wild P_aO_2 fluctuations to prevent RoP (below).

Tests: CXR; U&E; infection screen; glucose; Ca^{2+} ; Mg^{2+} . *Treatment:* If aspiration is the problem, give small feeds every 2h, or continuous tube feeds. Monitor $S_{A}O_2$ continuously; if hypoxic despite an ambient O_2 of 40%, CPAP or IPPV is needed. Caffeine citrate 10mg/kg (caffeine base) iv, then 2.5 mg/kg/day po/ivi helps and has fewer SEs than theophyllines. $n=85$ medline:10853619 & 11220405 Nasal CPAP \pm doxapram may prevent hypoventilation. If apnoea is seizure-related, see p 238. *Weaning:* Try 4–5 days after apnoea has stopped. *Prevention:* Betamethasone intrauterine maturation (p 102). medline:11085344

RoP (retinopathy of prematurity) Retinal vessels in prems react badly to fluctuating P_aO_2 , eg during resuscitations \rightarrow retinal fibrosis \rightarrow detachment \rightarrow vision \downarrow . *Prevalence (lower limits):* <1000g: 53%; $\leq 1250g$: 43%; $\leq 1500g$: 35%. medline:9923866

Classification: There are 5 stages, depending on site involved, the degree of retinal detachment, and extent (measured as clock hours in each eye).

Treatment: Diode laser therapy causes less myopia than cryotherapy.

Screening: If <1500g or ≤ 31 weeks' gestation. If ≤ 25 weeks, screen ophthalmoscopically under 0.5% cyclopentolate pupil dilatation at 6 weeks old, and 2-weekly until 36 postmenstrual weeks. Often one examination is enough if 26–31 weeks' gestation. It must be done by an experienced ophthalmologist.

IVH (intraventricular haemorrhage) occurs in 40% if birth wt $\leq 1500g$. *Risk factors:* $P_aO_2 \downarrow$, $P_aCO_2 \uparrow$, BP \downarrow , bleeding disorder (p 244), birth trauma, Caesarean section, Apgar <4 at 1min or <8 at 5mins, low birth weight, respiratory distress, PDA, anaemia, and umbilical arterial catheterization. medline:10996925

Suspect in any neonate who deteriorates rapidly (esp. in 1st week of life). Opisthotonos, cerebral irritability, shock, \downarrow feeding skills, a bulging fontanelle with a rapidly expanding head, an exaggerated Moro reflex, fits, and somnolence are telling signs, but ~50% may be 'silent'. *Tests:* Ultrasound; CT. *Complications:* IQ \downarrow , cerebral palsy, hydrocephalus. Many survive unscathed. It is uncertain if late learning and behavioural disorders occur. *Treatment:* Rest, head elevation, seizure control (p 238), surfactant (p 240). medline:11394231

UK randomized trial of extracorporeal membrane oxygenation (ECMO)
 This important trial ($N=185$ neonates of ≥ 35 weeks' gestation) showed that ECMO done after referral to one of the five UK ECMO centres reduced deaths from severe respiratory failure (oxygenation index ≥ 40). Typical diagnoses were pulmonary hypertension from meconium aspiration, diaphragmatic hernia, persistent fetal circulation, sepsis, and respiratory distress syndrome. The trial had to be stopped early because independent scrutineers found the results so strongly favoured ECMO over traditional measures: 30 out of 93 died in the ECMO group, compared with 54 out of 92 in the control group. The advantage was upheld irrespective of the primary diagnosis, or the type of referring centre. The number of infants suffering severe disability at follow-up at one year was one in each group.

$N=185$
 medline:8676720

ECMO is a complex procedure providing life-support in specialized units during respiratory failure, which obviates the need for lung gas exchange.

Criteria which may make ECMO cost-effective: weight $>2\text{kg}$; no major congenital malformations; no major CNS abnormality on ultrasound; gestation >34 weeks; oxygen index (OI) >40 (unresponsive to nitric oxide inhalation).

NB: more premature babies may benefit, eg when problems with circuitry and heparinization are solved. Need for ECMO may be $\sim 1/4000$ live births. It is thought to be economically worthwhile.

medline~9756807 &: 11126261

Ventilatory support for neonates

This is a skill to be learned at the cot side. Nurses are often expert, and may be best placed to teach. Needs of apparently very similar babies vary, so what follows is only a guide to prepare your mind before teaching. Continuous refinement in the light of transcutaneous and blood gas analysis is needed.

PTCCF (pressure-limited, time-cycled continuous flow ventilation) A continuous flow of heated & humidified gas is passed via an endotracheal tube. Nasotracheal siting may be best (fewer tube displacements), but insertion needs skill. medline:10796391 **Variables:** Air/O₂ mix; peak inspiratory pressure (PIP); positive end-expiratory pressure (PEEP); inspiratory (T_I) and expiratory (T_E) times. The infant can make respiratory efforts between ventilator breaths (intermittent mandatory ventilation, IMV). Aim for a T_I of 0.3–0.45sec.

Initial settings: Choose to give good chest inflation and air entry on auscultation and adequate transcutaneous O₂ readings. Typical settings might be T_I 1sec, 20–25 cycles/min, inspiratory pressure 14–16cmH₂O, and PEEP 5cmH₂O. Adjust in the light of blood gas analysis.

PEEP (positive end-expiratory pressure) A loaded valve is fitted to the expiratory limb of the ventilator, so airways pressure stays ≥atmosphere. Levels >10cmH₂O are rarely used (venous return to the thorax ↓).

CPAP (continuous positive airways pressure) Pressure is raised throughout the respiratory cycle, so assisting spontaneous inspiration. With skill, this method has few complications, and is useful as a first stage in ventilating a baby before it is known whether he will need IMV.

PTV (patient-triggered ventilation) Babies tend to fight ventilators, trying to expire during inflations: hence the need for paralysis: but this makes them dependent on the ventilator, fluid retention is common, and limb contractions may develop—also air-trapping, ↓gas-exchange, pneumothorax and bronchopulmonary dysplasia. Paralysis helps here by ↓rate of pneumothorax and ivh (p 232). Another option is PTV: here inspiratory and end-expiratory pressure is set by the operator, but the rate set by the baby. PTV is associated with a shorter duration of ventilation. medline:11279692 & 10963241 Hiccups cause problems if abdominal movement is used to detect inspiration.

SIMV (synchronous intermittent mandatory ventilation), **PTV** (above) & **HFOV** (high frequency oscillatory ventilation) are nonstandard methods. medline:11279692

Paralysis Pancuronium (eg 30µg/kg iv; then 10µg/kg every 1.5–4h to maintain paralysis) prevents pneumothorax ± ivh (p 232) in asynchronous respiratory efforts (eg needing unexpectedly high P_I). medline:11034760

Other factors **Pain relief:** Consider 5% dextrose ivi with morphine 100–200µg/kg over 30min, then 5–20µg/kg/h. This is thought safe, and ↓catecholamine concentrations (a possible objective correlate of pain/stress).

Air leak: Air ruptures alveoli tracking along vessels and bronchioles (pulmonary interstitial emphysema), and may extend intrapleurally (pneumothorax + lung collapse), or into the mediastinum or peritoneum. It is less common with HFOV. **Signs:** Tachypnoea, cyanosis, chest asymmetry. The lateral decubitus cxr is often diagnostic. Prompt 'blind' needle aspiration of a pneumothorax may be needed. Aspirate through the second intercostal space in the midclavicular line with a 25G 'butterfly' needle and a 50mL syringe on a 3-way tap. If the leak is continuous, use underwater seal drainage.

Weaning ↓Rate of IMV & lower P_I by ~2cm H₂O decrements; try extubation if blood gas ok with ~4cmH₂O PEEP with spontaneous breaths. **NB:** racemic adrenaline doesn't stop intubation-associated ↑upper airways resistance. medline:11279697

Bronchopulmonary dysplasia (BPD)

BPD (*chronic lung disease of prematurity*) is a big complication of ventilation for respiratory distress syndrome. There is persistent hypoxaemia \pm difficult ventilator weaning. Classically, BPD is mainly from barotrauma and oxygen toxicity, whereas surfactant-related BPD is multifactorial with airway infections triggering inflammatory cascades. (Without surfactant, many would not survive to get BPD.) 2001 medline:11359038 Oxidative processes may also play a key rôle, but antioxidants are unproven. 2001 medline:11281247

Tests: *cxr*: hyperinflation, rounded, radiolucent areas, alternating with thinner strands of radiodensity. *Histology*: necrotizing bronchiolitis + alveolar fibrosis.

Mortality: Variable, \therefore complex interaction with surfactant use. medline:11359038

Early sequelae: Decreased cognition, cerebral palsy, feeding problems. O_2 desaturation during feeds is not uncommon.

Late sequelae: By adolescence/early adulthood the main changes remaining are airways obstruction, airways hyper-reactivity, and hyperinflation.

Prevention: Antenatal steroids + postnatal surfactant treatment. If protracted mechanical ventilation is necessary, low doses of iv glucocorticoid steroids in the 2nd & 3rd week of life are administered, together with bronchodilators and 'suitably high' calorie feeding. medline:11328969

Pulmonary hypoplasia

Suspect this in all infants with persisting neonatal tachypnoea \pm feeding difficulties, particularly if prenatal oligohydramnios. Hypoplasia may be a consequence of oligohydramnios, eg in Potter's syndrome or premature rupture of the membranes. In diaphragmatic hernia it is a consequence of the 'space-occupying lesion'. Hypoplasia may also be a primary developmental abnormality. Diagnosis may be prenatal, 2000 medline:11169336 postmortem, or, if facilities are available, by measuring functional residual capacity. *cxr* is likely to be misleadingly reported as normal. The condition need not be fatal: postnatal catch-up growth has been well documented.

Differential diagnosis: Respiratory distress syndrome if premature, or meconium aspiration, sepsis, or primary pulmonary hypertension. medline:10836158

Some degree of pulmonary hypoplasia is the price of adopting an expectant management plan for early spontaneous rupture of the membranes, but despite this, expectant management leads to fewer deaths. medline:10871491

Is he/she too premature for active treatment?

Taking into account gestation, condition at birth, parental expectations, ethical considerations, tendencies to paternalism, and medical economics is difficult, which is why some centres have an arbitrary cut-off for gestations before eg 25 weeks (eg in Leiden, Holland). The Royal College of Paediatricians view is that each baby merits individual consideration. Don't expect to get all these decisions right—but, equally, don't stop trying.

Neonatal jaundice

By allowing the early management of jaundice, transcutaneous bilirubin levels measured by midwives in homes may prevent kernicterus in babies discharged early. In non-Caucasians, the device needs recalibration: don't rely on tests such as pressing the nose. medline:11275655

Hyperbilirubinaemia ($<200\mu\text{mol/L}$) after the 1st day of life is common and may be 'physiological' (eg in binding free radicals). Mechanisms:

- 1 Hepatic immaturity in bilirubin conjugation \pm \uparrow removal and destruction of fetal RBCs. If albumin \downarrow unconjugated bilirubin is left unbound.
- 2 Absence of gut flora impedes elimination of bile pigment.
- 3 \downarrow Fluid intake; breast feeding (not *usually* a reason to stop). medline:11331689

Jaundice within 24h of birth is always abnormal. *Causes:* ►Sepsis or:

- **Rhesus haemolytic disease:** +ve direct Coombs' test (DCT, p 242).
- **ABO incompatibility:** (mother O; baby A or B, or mother A and baby B, or vice versa) DCT +ve in 4%; indirect Coombs +ve in 8%. Maternal IgG anti-A or anti-B haemolysis is 'always' present. medline:10830015

High-dose iv immunoglobulin is one therapy to consider. medline:10102158

- **Red cell anomalies:** congenital spherocytosis (do blood film); glucose-6-phosphate dehydrogenase deficiency (do enzyme test). medline:11317924

Tests: FBC; film; blood groups (eg rare group incompatibility.); Coombs' test; urine for reducing agents; syphilis/TORCH^{et al} screen, p 98. medline:9365074

Prolonged jaundice (not fading after 9 days) *Causes:* breast feeding; sepsis (UTI & TORCH, p 98); hypothyroidism; cystic fibrosis; biliary atresia (esp. if jaundiced for >14 days): look for *conjugated* bilirubin and pale stools.

Galactosaemia: urine tests for reducing agents (eg Clinitest®) are +ve, but specific tests (Clinistix®) for glycosuria are -ve (an insensitive test; galactose-1-phosphate uracyl transferase levels are diagnostic).

Kernicterus May occur if bilirubin is $>360\mu\text{mol/L}$ (lower in prems).

Signs: sleepy \pm poor suck (stage I), through to 'setting sun' lid retraction, odd movements, cerebral palsy, deafness and IQ \downarrow (later, and stage IV). Lesser levels ($170\text{--}323\mu\text{mol/L}$) are unlikely to cause permanent problems, and have little effect on IQ unless preterm or light-for-dates. It is prevented by phototherapy (below) \pm exchange transfusion. medline:11519982

Starting phototherapy A *plasma bilirubin guide:* (use your units protocol): *Term baby at age 48h:* $230\mu\text{mol/L}$. (At 72h $250\mu\text{mol/L}$; at 4 days $275\mu\text{mol/L}$; at 5 days $300\mu\text{mol/L}$.) If prem (but >25 weeks) or low-birth-weight, \downarrow thresholds by $\geq 25\mu\text{mol/L}$. Stop when levels fall (eg by $>25\mu\text{mol/L}$) below these thresholds. SE: $T^\circ \uparrow \downarrow$; eye damage; diarrhoea; separation from mother; fluid loss (give 30mL/kg/day extra water). *Intense phototherapy* is an adjunct to exchange transfusion. medline:9307164 & 8695979 & 2782930

Exchange transfusion Uses warmed blood (37°C), crossmatched vs maternal serum, given ideally via umbilical vein iv. Aim to exchange 160mL/kg over $\sim 2\text{h}$ (\approx blood volume $\times 2$). iv Ca^{2+} isn't needed if citrate phosphate dextrose blood is used. Monitor ECG, U&E, Ca^{2+} , bilirubin & glucose. Consider more exchanges if bilirubin goes on rising. *Stop* if the pulse rate fluctuates by >20 beats/min. ►Ensure the volumes exchanged always balance. If anaemic, consider a simple fresh blood transfusion (20mL/kg). *Complications:* Pulse \downarrow , apnoea, platelets \downarrow , glucose \downarrow , Na^+ \downarrow . medline:8682393

Bilirubins needing exchange transfusion: *Term baby at birth,* $50\mu\text{mol/L}$; at 12h, $125\mu\text{mol/L}$; at 24h, $200\mu\text{mol/L}$; at 36h, $250\mu\text{mol/L}$; at 48h, $325\mu\text{mol/L}$; at 72h, $350\mu\text{mol/L}$; at 4 days, $375\mu\text{mol/L}$; at 5 days, $400\mu\text{mol/L}$. If premature or weight <2.5 kg lower these thresholds (eg by $50\mu\text{mol/L}$, after age 48h).

►► The ill neonate

Sepsis Common (1–10/1000 births), and commonly overwhelming (mortality 15–50%). Here, signs may be minimal, or as OPPOSITE.

Action: Clear the airway; intubate and ventilate if necessary. This will correct acidosis, so bicarbonate is rarely needed. Set up a colloid iv (20mL/kg initially). Exclude hypoglycaemia; do blood gases.

Infection screen: •Blood-culture, virology, FBC, platelets, glucose, CXR.

•Lumbar puncture: csf (p 258) for urgent Gram stain, cell count, protein & glucose level, culture, and virology. •Stool: for virology.

•Urine: microscopy, culture, and virology. •ENT swabs: for culture.

Antibiotics: In early onset infection (<48h old) group B streps and *E coli* (+ any other organism prevalent in your NICU/SCBU) are common. Benzylpenicillin 25mg/kg/12h iv (per 8h if >7 days old; per 6h if >1 month) + netilmicin 6mg/kg/24h iv (5mg/kg in prems <35 weeks' gestation).

Notes: IM benzylpenicillin: dilute 600mg in 1.6mL water; 600mg of the powder displaces 0.4mL, so final concentration is 300mg/mL.

In late onset infection (>48h old), coagulase -ve staphs are a likely cause, so regimens include vancomycin or teicoplanin (p 191). *Staph aureus* is rarer—flucloxacillin dose: 50–100mg per 12h iv over 3min (per 8h if 7–21 days old; per 6h if >21 days old; If creatinine clearance <10mL/min/1.73m², ↑ the dosage interval. Alternative: cefotaxime, p 258.

If the csf suggests meningitis, see p 258.

Neonatal seizures (~4/1000 births) **Causes:** P₅O₂↓; infection; glucose↓, CNS damage (haemorrhage, hydrocephalus, CNS cysts); Ca²⁺ <1.7mmol/L; Na⁺↑↓; Mg²⁺↓ (eg <0.4mmol/L, see below).

medline:11358751

Treatment: Turn on the side. Ask an experienced nurse to help.

•Rule out (or treat) hypoglycaemia (5mL/kg 10% dextrose iv over 5min, then 100mL/kg/day ivi + hourly feeds, ↓ivi rate as plasma glucose allows).

•Phenobarbital 15mg/kg iv over 5mins + 5mg/kg at 1h if needed. Maintenance: reduce slowly from 2mg/kg/12h iv/po. Monitor blood levels. Aim for 30–45μg/mL >4h after a po dose; t_{1/2}=3 days at first, so once daily doses are used; t_{1/2} halves after 10 days as liver enzymes are induced.

•If seizures continue, give phenytoin 20mg/kg ivi in 0.9% saline (<1mg/kg/min; aim for a plasma level of 10–20μg/mL >1h post dose.).

Other agents: clonazepam; lorazepam; paraldehyde).

•If fits continue, try pyridoxine ≤100mg iv (then daily po); monitor EEG.

Hypocalcaemia: Use low PO₄³⁻ milk + Ca gluconate 10%, 4.4mL/kg/day po (iv: 0.3mL/kg diluted in 4.8mL/kg of saline over 10min). Monitor ECG. NB: some favour higher iv emergency doses, eg 2mL/kg of the 10% solution).

Hypomagnesaemia: Give MgSO₄ 10% (100mg/mL), eg 100mg/kg iv.

Shock Causes: Blood loss (placental haemorrhage, twin–twin transfusion; intraventricular haemorrhage; lung haemorrhage); capillary plasma leaks (sepsis, hypoxia, acidosis, necrotizing enterocolitis); fluid loss (p&v; ↑insensible loss; inappropriate diuresis); cardiac causes (hypoxia, hypoglycaemia, L to R shunts, valve disease, coarctation). **Signs:** Pulse ↑; BP ↓; urine output ↓; coma. **Management:** Ventilate as needed. Treat causes. Aim for a cvp of 5–8cmH₂O. Give colloid 10–20mL/kg iv over 30–60min, with dobutamine 2–15μg/kg/min (up to 40μg/kg/min in older children) as needed (which may be combined with dopamine for synergism: 2μg/kg/min; increase by 2μg/kg/min increments to 20μg/kg per min). If the pH is <7.2, bicarb, eg 1–2mmol/kg iv as a 4.2% solution, may be indicated (once adequate ventilation is established; don't mix with inotropes).

Problems with neonatal sepsis

Diagnosis is often hard, as signs may be nonspecific, and cultures take time—and time is most definitely *not* on your side. Signs include:

- Unusual crying ● Sleepiness ● Listlessness ● Shock ● Fits/apnoea
- Hypotonia ● Vomiting ● Rashes ● T° ↑ or ↓ ● Bradycardia

Also: feeding difficulty, abdominal wall reddening (omphalitis), grunting, rib recession, ↑ use of respiratory muscles, tachypnoea, cyanosis, cool peripheries; ↓ capillary return on pressing the skin.

Nonspecific and probably unreliable tests include C-reactive protein, FBC & film: looking at the ratio of immature to total neutrophils *may* help.

NB: it's possible that looking for soluble immunological mediators may help: ideal ones may be interleukin-1 receptor antagonist (IL-1ra), and interleukin-6 (IL-6). Specificity: 92% and 83%, respectively. Circulating intercellular adhesion molecule-1 (ICAM-1) is less reliable. medline:9788457

Blood and other cultures are definitive, but take ~48–72h.

Normal values are often hard to define—eg a csf wcc of up to 30/mm³ with 60% polymorphs may be normal in neonates.

Changing patterns of antibiotic resistance: For example, resistance to netilmicin varies from 36% to 11% of isolates. medline:10565074 & 10225325
In hospital-acquired infections, amikacin may be needed. medline:9722468

Drug pharmacokinetic problems: These are only partly obviated by doing levels. Creatinine, sex, and birthweight influence what is an acceptable level in complex ways. With netilmicin, adjust dose as indicated in the *Data sheet* if renal function is poor. Aim for a 1h post-dose (peak) level of 12mg/L, and ≤2mg/L at 14–18h. If levels are problematic, cefotaxime is a good alternative (p 258). medline:10872642

Subtle manifestations of neonatal seizures

Unless you have a high index of suspicion, you may miss seizures referable to the brainstem, eg nystagmus, conjugate eye movements, posturing, sucking movements, lip smacking, etc. Grand mal is rare. EEGs are helpful in diagnosing when unsure. Always look for metabolic abnormalities (these are treated with metabolic approaches; don't rely on conventional anticonvulsants).

2001 medline: 11282039

Respiratory distress syndrome (RDS)

Insufficient surfactant is made so that the lungs are unable to stay expanded; re-inflation between breaths exhausts the baby, and respiratory failure follows. It is the major cause of death from prematurity.

Infants at risk: 100% if 24–28 weeks' gestation; 50% if 32 weeks. Also: maternal diabetes, male sex, second twin, Caesarean birth.

Prenatal prevention See p 102.

Signs Worsening tachypnoea ($>60/\text{min}$) in the first 4 hours after birth. Increased inspiratory effort, with grunting, flaring of the nasal alae, intercostal recession and cyanosis. CXR: diffuse granular patterns, with air bronchograms. Mild signs subside after ~36h.

Differential diagnosis *Transient tachypnoea of the newborn* is due to excess lung fluid. It usually resolves after 24h. *Meconium aspiration* (p 244); congenital pneumonia (group B streptococci); tracheo-oesophageal fistula (suspect if respiratory problems after feeds); congenital lung abnormality.

Treatment Learn at the cot side. If gestation ≤ 28 weeks, intubate at birth and, if $>700\text{g}$, give surfactant by ET tube, eg bovine Beractant[®], 4mL/kg within 8h of birth \pm 2nd dose in 6h, or at 2h if needed. 2001 medline:11332171
Rock gently to aid spread in the bronchial tree; max number of doses: 4. Monitor O_2 as needs may suddenly \downarrow . medline:10791521

- Wrap up warmly and take to NICU/SCBU incubator.
- Monitor blood gases (eg transcutaneously). Aim for a $P_a\text{O}_2$ of 7–12kPa, enhancing the ambient O_2 with a perspex head box.
- If blood gases worsen, intubate and support ventilation (start with CPAP, p 234), before exhaustion sets in. A rising $P_a\text{CO}_2$ may indicate either that CPAP is too high, or that paralysis and further ventilation is needed (CPAP = continuous positive airways pressure).
- *Traditional ventilator settings*: See p 234. Inspiratory pressure 20cmH₂O with 60% O_2 ; positive end-expiratory pressure 3–5cmH₂O; 20–25 breaths/min, with an inspiratory duration of 1sec. On connecting the endotracheal tube check chest movement is adequate and symmetrical. Listen for breath sounds. $P_a\text{O}_2$ is increased by \uparrow pressures (not too high). $P_a\text{CO}_2$ is decreased by \uparrow breath frequency by lessening inspiratory time. One option needing further verification is *high-frequency oscillatory ventilation*. cochrane medline:10796129 & 10828581
- \blacktriangleright If any deterioration, consider: blocked or dislodged tube (a common occurrence), infection, faulty ventilator, pneumothorax.
- *Fluids*: Avoid milk for 24–28h. Give 10% dextrose ivl (p 246). *Nutrition*: Get expert help. Inositol is an essential nutrient promoting surfactant maturation and may play a vital rôle in neonatal life. Supplementing nutrition of prems with inositol can reduce RDS complications. medline:11034685 & 2117207

Signs of a poor prognosis Persistent pulmonary hypertension, large right to left shunt via the ductus; \uparrow dead-space fraction in lungs. Nutton NEJM 2002 346 1281

If, despite everything, hypoxia worsens, the baby is dying. Confer with a senior colleague. Explain what is happening to the mother, and that the baby will feel no pain. Encourage christening, or whatever is congruent with the parents' beliefs. Give pain relief (p 198). Disconnect the tubes, so allowing the mother to hold the baby, and, in so doing, to aid her grief.*

*Roy Col Paed & Child Health 1997 *Withholding or Withdrawing Life-saving Treatment in Children*.

Communicating with parents

It is very important to take time to explain to parents exactly what is happening to their baby—not just for the respiratory distress syndrome, but for *any* serious diseases. Structured, tested interviews yield these guidelines:

medline:2503155

- Ask both parents to be present (plus a nurse whom they trust).
- Take steps to avoid being interrupted. Allow sufficient time.
- Call the parents by name.
- Do not avoid looking at the parents (mutual gaze promotes trust).
- Name the illness concerned with its complications. Write it down.
- Outline the support available throughout the illness.*
- Elicit what the parents now know. Clarify or repeat as needed.
- Answer any questions. Acknowledge that it is difficult to absorb all the information so arrange early follow-up.

*Give the address and phone number if there is a patient's group to help. NB a central directory exists for many neonatal and other conditions. Access these by phoning a central directory of groups (in the UK the number is: 020 7240 0671). www.grief.com/Children.htm

Rhesus haemolytic disease

Physiology When a RhD-ve mother delivers a RhD+ve baby a leak of fetal red cells into her circulation may stimulate her to produce anti-D IgG antibodies (isoimmunization). In later pregnancies this can cross the placenta, causing worsening rhesus haemolytic disease (*erythroblastosis fetalis*) with each successive Rh+ve pregnancy. First pregnancies may be affected due to leaks, eg: ●Threatened miscarriage ●APH ●Mild trauma ●Amniocentesis ●Chorionic villous sampling ●External cephalic version

An affected oedematous fetus (with stiff, oedematous lungs) is called a *hydrops fetalis*. Anaemia-associated ccf causes oedema, as does hypoalbuminaemia (the liver is preoccupied by producing new RBCs). $\Delta\Delta$: Thalassaemia; infection (eg toxoplasmosis, CMV, p 98); maternal diabetes.

Clinical Rh disease ►Test for D antibodies in all Rh-ve mothers, at booking, 28 & 34 weeks' gestation. Anti-D titres <4u/mL (<1:16) are very unlikely to cause serious disease; it is wise to check maternal blood every 2 weeks. If >10u/mL, get the advice of a referral centre: fetal blood sampling \pm intraperitoneal (or, with fetoscopy, intravascular via the cord) transfusion may be needed. Expect fetal Hb to be <7g/dL in 10% of those with titres of 10–100u/mL (75% if titres >100u/mL). ^{1, 2, 14} medline:8935259

Do regular ultrasound (+amniocentesis if anti-D titre >4u/mL). Timing is vital. Do it 10 weeks before a Rh-related event in the last pregnancy (eg if last baby needed delivery at 36 weeks, expect to do amniocentesis at 26 weeks). Fetuses tolerating high bilirubins may be saved risky transfusions (fatality 2–30%) if monitored by serial measurements of fetal Hb (by fetoscopy or non-invasive middle cerebral artery peak velocity) and daily ultrasound, to detect oedema, cardiomegaly, pericardial effusion, hepatosplenomegaly, or ascites. medline:10620643

Anti-D is the chief antibody. Others: Rh C, E, c, e, Kell, Kidd, Duffy (all are IgG). Low concentrations sometimes produce severe disease.

Prognosis is improving. Mortality is <20% even for hydropic babies. Note that maternal antibodies persist for some months, and continue to cause haemolysis during early life. medline:9128296

Signs (For *kernicterus*, see p 236.)

- Jaundice on day 1 (or later)
- Heart failure (oedema, ascites)
- Progressive anaemia; bleeding
- Yellow vernix
- Hepatosplenomegaly
- CNS signs

Tests FBC (Hb↓), reticulocytes and direct Coombs test,* for all patients.

- Anti-Rh agglutinins present
- Mother Rh-ve, baby Rh+ve
- Anti-Rh titre ↑ in mother
- Reticulocytes ↑
- Bilirubin ↑
- Hypoglycaemia

Exchange transfusion *Indications/technique*: p 236. If Hb <7g/dL, give 1st volume of the exchange transfusion (80mL/kg) as packed cells, and subsequent exact exchanges according to response. ►Keep the baby warm.

Ultraviolet photodegradation of bilirubin (with phototherapy lamp) may be all that is needed in less-than-severe disease. Give extra water (30mL/kg/24h po). Avoid heat loss. Protect the eyes.

Giving Rh-ve mothers anti-D immunoglobulin (p 109) This strategy has markedly reduced need for exchange transfusion. medline:10485296

ABO incompatibility 1 in 45 of group A or B babies born to group O mothers will have haemolysis from maternal antibodies. Exchange transfusion may be needed, even in first-borns.

Hydrops fetalis: management

- Get expert help.
- At birth, take cord blood for Hb, PCV, bilirubin (conjugated and unconjugated), blood group, Coombs' test,* serum protein, LFTs, and infection screen (p 98) to find the cause—eg isoimmunization; thalassaemia; infection (eg toxoplasmosis, syphilis, parvoviruses, CMV, p 98); maternal diabetes; twin–twin transfusion; or hypoproteinaemia.
- Expect to ventilate with high inspiratory peak pressure and positive end pressure.
- Monitor plasma glucose 2–4 hourly, treating any hypoglycaemia.
- Drain ascites and pleural effusions if severe.
- Correct anaemia.
- Vitamin K 1mg, to reduce risk of haemorrhage.
- If ccf is present, furosemide may be needed, eg 1–2mg/kg/12h iv.
- Limit iv fluids to 60mL/kg/24h (crystalloid); if exchange transfusing, aim for a deficit of 10–20mL/kg. Monitor urine output.
- Prognosis: 90% of those with non-immune hydrops die *in utero*; 50% die postnatally. Babies with non-immune hydrops not secondary to infection have a good neurological outcome.

2000 ^{BJOG} medline:10688515

*The direct Coombs' test identifies red cells coated with antibody or complement and a positive result usually indicates an immune cause of haemolysis (OHCM p 638).

Other neonatal problems

Necrotizing enterocolitis (NEC) This is necrosis of bowel mucosa. Typically it occurs at the end of the first week of life in a prem infant on NICU/SCBU (it affects 1–2% of admissions to NICU). If mild, a little blood and mucus may be passed per rectum. At worst, there is sudden abdominal distension, tenderness (\pm perforation), shock, DIC, and sloughing of the rectal mucosa. It may be sporadic or epidemic. **Treatment:** **Stop oral feeding**; barrier nurse; culture faeces; do erect and supine abdominal x-rays, to look for oedematous loops of bowel with intramural gas. Crossmatch blood (to correct anaemia); give antibiotics—metronidazole 7.5mg/kg/12h iv with penicillin and netilmicin (p 238) and regimens of cefotaxime with vancomycin have been used. medline:3680079

Liaise early with a surgeon. Do repeated imaging and girth measurement. Platelets mirror disease activity; $<100 \times 10^9/L$ is 'severe'. 2001 medline:11329593

Laparotomy indications: progressive distension, perforation. **Prophylaxis:** Oral antibiotics may (perhaps) reduce incidence of NEC (NNT \approx 10). cochrane medline:11279690

Meconium aspiration syndrome Only thick meconium is important (aspiration pneumonitis). Suck out pharynx and trachea with oral mucus extractor after delivery of the head, but before the body is born. If meconium is present, intubate the trachea (large tube) for suction. The obstetrician may try compressing the thorax as he passes the baby to the paediatrician, so minimizing inspiratory effort until the airway is clear (this doesn't stop diaphragm effort). Observe closely, postnatally. CPAP or IMV may be needed (p 234). Try a PEEP of 6cm H₂O and an inspiratory time of 0.5–0.75sec. Give penicillin + netilmicin (p 238). Use of anti-inflammatories, high-frequency ventilation, exogenous surfactant, inhaled nitric oxide and liquid ventilation are unsupported by randomized evidence. medline:11332953

Haemorrhagic disease of the newborn occurs from days 2–7 postpartum; it is due to lack of vitamin K (no enteric bacteria, or mother taking anticonvulsants). The baby is usually well, apart from bruising/bleeding. Prothrombin & partial thromboplastin times (PT & PTT) \uparrow ; platelets normal. **Prevention:** (many regimens) eg vit K eg 2mg IM (if at \uparrow risk) \pm oral vit K if at low risk. \blacktriangleright Formula milk already contains vit K. **Treatment:** Plasma (10mL/kg iv) and vit K (eg 1mg iv) for active bleeding (monitor coagulation). 2001 medline:11348908

Disseminated intravascular coagulation (DIC) **Signs:** Septic signs (ill), petechiae, venepuncture oozings; GI bleeding. **Tests:** Platelets \downarrow , schistocytes (fragmented RBCs); INR \uparrow , fibrinogen \downarrow ; partial thromboplastin time \uparrow ; D-dimer \uparrow (hard to interpret if birthweight very low). medline:9834015 & 9369697

Treatment: Get help; treat cause (eg infection, NEC); give vit K 1mg iv; then:

- Platelet transfusion, to keep platelets $>30 \times 10^9/L$.
- Fresh plasma, eg 10mL/kg iv \pm heparin iv \pm protein C. medline:10632474
- If bleeding still continues, consider exchange transfusion.

Immune thrombocytopenia $<10\%$ of babies of women with autoimmune thrombocytopenia (ITP) get thrombocytopenic (p 142). **Alloimmune thrombocytopenia:** (1:2000 births; via fetomaternal incompatibility of platelet antigens). It develops *in utero*. 50% are 1st born (it recurs in $\sim 80\%$ of later pregnancies with same or \uparrow severity). If affected *in utero* 25% have CNS problems; 10% an intracerebral haemorrhage *in utero*. Platelets continue to fall for 48h post-delivery. Treat severe thrombocytopenia with *compatible platelets* or *irradiated maternal platelets*. IgG 1g/kg/24h iv for ~ 2 days may help. Platelet transfusion via cordocentesis from 24wks may be needed in later pregnancies. Diagnose by detecting *maternal platelet alloantibody* against father's platelets. Do *neuroimaging* on all patients. medline:10637839

Minor neonatal problems

Most neonates have a few insignificant lesions; the more you examine neonates the better you will become at reassuring mothers. medline:1870904

Milia: These tiny cream sebaceous cysts on the nose or palate (Epstein's pearls) are self-limiting. Differential: pyogenic granuloma. medline:11169281

Erythema toxicum (neonatal urticaria): These are harmless red blotches, often with a central white vesicle which come and go in crops. They last ~24h (try ringing them), in contrast to septic spots which develop obvious pustules. If in doubt take a swab. Ask for a Wright's stain: neutrophils indicate sepsis; eosinophils suggest erythema toxicum. medline:10370809

Miliarial/heat rash: This itchy red rash fades rapidly when the baby is unwrapped. It is found in >8% of babies in some populations. medline:3952030

Stork mark: These are areas of capillary dilatation on the eyelids, central forehead and back of the neck—where the baby is deemed to have been held in the stork's beak. They blanch on pressure and fade with time.

Harlequin colour change: One side of the face or body suddenly flushes for a few minutes. It is a shortlived vasomotor event.

Peeling skin: Common in postmature babies, it does not denote future skin problems. Olive oil prevents skin folds from cracking.

Petechial haemorrhages, facial cyanosis, subconjunctival haemorrhages: These temporary features generally reflect suffusion of the face during delivery (sometimes inaccurately referred to as 'traumatic asphyxia').

Swollen breasts: These occur in both sexes and occasionally lactate (witch's milk). They are due to maternal hormones and gradually subside if left alone, but if infected require antibiotics. Witch's milk may persist until 2 months of age. medline:3946357

The umbilicus: It dries and separates through a moist base at about day 7. Signs of infection: odour, pus, periumbilical red flare, malaise. Isolate the baby, take swabs and blood cultures, give antibiotics. Granuloma: exclude a patent urachus and cauterize with a silver nitrate stick

Sticky eye: (Common; usually from an unopened tear duct, p 482.) Swab to exclude *ophthalmia neonatorum* (p 100)/chlamydia (special swab).

Feeding anxieties: Healthy term babies require little milk for the first few days and early poor feeding is not an indication for investigation or bottle top-ups. The exceptions are babies of diabetic mothers, and light-for-dates babies, because of their risk of hypoglycaemia.

New babies may have difficulty co-ordinating feeding and breathing and briefly choke, gag, or turn blue. Exclude disease, check feeding technique (too much? too fast?) and reassure.

Regurgitation is often due to overfilling a tiny stomach with milk and air. Check feeding technique; if bottle fed, is the teat too big for the mouth or the hole too small or the amount too great?

Winding during feeds may help but is not essential to health.

Red-stained nappy: This is usually due to urinary urates but may be blood from the cord or baby's vagina (oestrogen withdrawal bleed).

Sneezing: Neonates sneeze to clear amniotic fluid from the nose.

Enteral and parenteral nutrition

Breastfeeding (p 176–8) is the ideal way to feed babies, but not all neonates can do this (and we must respect mothers who choose not to breastfeed). Here, formula milk is a viable alternative (p 179), as is expressed breast milk (EBM—technique: p 177). EBM is often inadequate to supply the increased demands of preterm infants (both calories and ion content). If <2kg, fortify with powder sachets of Breast Milk Fortifiers® (Cow & Gate).

Gavage tube feeding *Indications:* Any sick infant who is too ill or too young to feed normally (eg respiratory distress syndrome). Expressed breast milk or formula milk is fed via a naso- or oro-gastric tube either as a bolus or as a continuous infusion.♦ If gastro-oesophageal reflux or aspiration is a problem, then a silastic naso-jejunal tube can be used. After entering the stomach, the tube enters the jejunum by peristalsis: confirm its position on x-ray. When the baby improves, start giving some feeds by mouth (PO), eg 1 PO per day, progressing to PO every 3rd feed. Then try alternating PO and gavage; then gavage every 3rd feed; finally try all PO. If during PO feeds, cyanosis, bradycardia or vomiting supervene, you may be trying too soon.

www.neonatology.org/syllabus/feeding.premature.html

Trophic feeding *Synonyms:* Minimal enteral feeding; gut priming; hypocaloric feeding. *Rationale:* If prems go for weeks with no oral nutrition, normal GI structure and function are lost despite an anabolic body state. Villi shorten, mucosal DNA is lost, and enzyme activity is less. Feeding a small volume of milk may prevent this.

Technique: Typically, milk volumes of ~1mL/kg/h are given by tube starting on day 2–3. The milk employed may be expressed breast milk or a preterm formula (eg Nutriprem®).

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Effects: Studies show that weight gain and head growth is improved, and that there are fewer episodes of neonatal sepsis, fewer days of parenteral nutrition are needed, and time to full oral feeding is less.

Eligibility: Experience shows that almost all prems with nonsurgical illness tolerate at least some milk as trophic feeds.

2000 medline~10634838

Parenteral nutrition (PN) This is given into a central vein. *Indications:* post-op; trauma; burns; if oral nutrition is poor (eg in ill, low-birth-weight babies) and necrotizing enterocolitis (when the gut must be 'rested'). *Day-by-day guide:* see box. ▶ *Sterility is vital;* prepare using laminar flow units. Monitoring needs to be meticulous.

Daily checks: Weight; fluid balance; u&e; blood glucose; Ca²⁺. Test for glycosuria. Change iv sets/filters. Culture filters, Vamin® & Intralipid® samples.

Weekly: Length and head circumference; skin fold thickness. LFTs; Mg²⁺; PO₄³⁻; alk phos; ammonia; triglycerides; FBC; ESR or CRP (helps determine if sepsis is present).

Complications: Infection; acidosis; metabolic imbalances; thrombophlebitis; hepatobiliary stenosis; cholelithiasis; osteopenia. medline:11132460 If plasma PO₄³⁻ ↓, consider giving PO₄³⁻ (0.25–0.5mmol/kg/day) as the potassium salt. Mix with dextrose, but not Vamin® or trace element mixtures. PN is complex: get expert help. In addition, some precipitation errors are preventable by using computer-based decision support.

medline:11079964

Stopping iv nutrition Do in stages to prevent hypoglycaemia.

Parenteral nutrition: day-by-day guide. ►All values are per kg/day.

Type of baby	Day of PN	Age days	PROTEIN	CARBO-HYDRATE	FAT	IONS (mmol)		FLUID		
			Vami-nolact [®] mL	Dextrose 10%; mL	Intralipid 20%; mL	Na	K	Ca	PO ₄	PN vol-ume; mL ¹
Neonates and low-birth weight babies	1	3	20	97.5	2.5	3	3	1	0.4-1	120
	2	4	30	115	5	3	3	1	0.4-1	150
	3	5	40	115	10	3	3	1	0.4-1	165
	M	>5	50	100	15	3	3	1	0.4-1	165
Infants	1		20	95-125	5	2.5	2.5	0.6	0.4	120-150*
>1 month	2		30	80-110	10	2.5	2.5	0.6	0.4	120-150*
& <10kg	M		40	65-125	15	2.5	2.5	0.6	0.4	120-150*
10-30kg	1&2		14	23.5-78.5	7.5	2	2	0.2	0.1	45-100*
	M		28	7-57	10-15	2	2	0.2	0.1	45-100*
>30kg	1&2		14	26-56	5	1.5-2	1.5-2	0.2	0.1	45-75*
	M		21	14-51.5	10-12.5	1.5-2	1.5-2	0.2	0.1	45-75*

Note: M=maintenance. *See p 250 for 24h fluid requirement.

¹ This is the total vol of fluid required (60 & 90mL/kg for days 1 & 2). *Trace elements:* Peditrace[®] eg 1mL/kg (max 10mL) if renal function OK—caution if prolonged treatment, eg >1month, or if ↓biliary excretion (stop if cholestasis); checking Mn⁺ may be needed. *Vitamins:* Solivito N[®]: add 1mL/kg/day (max 10mL) to Vaminolact[®] (protect from light). Cater for A, D, & K with Vitalip N[®] 1-4mL/kg/day (max 10mL/kg/day) to Intralipid[®]. ►These values are a guide only. Individual needs vary greatly. Get expert help.

►►The ill and feverish child

There are 2 distinct questions to ask: *How severe are the symptoms and How appropriate is the child's response to the illness?* The symptom may be severe (eg 'terrible diarrhoea all over the cot') without being biologically serious (if the baby is alert, drinking, wetting many nappies, and behaving as usual); but ANY apparently mild symptom should set your internal alarm bell ringing if:

medline:2201263 &: 1437423

- <Half the usual amount of feed has been taken in the last day.
- There is breathing difficulty, or high-pitched continuous moans or cries.
- There is a history of being pale, mottled, cyanosed, and hot.
- Dull expression; apathetic; uninterested in you; drowsy; dehydrated.
- Fewer than 4 wet nappies in the last 24h. • Blood in diarrhoea. • Seizures

Observe the limbs: pain causes *pseudoparalysis*, eg with legs in a 'frog position' (hips & knees semiflexed; feet rotated outward), eg in osteomyelitis, septic arthritis, syphilis, CMV, or scurvy).

medline:2211769 &: 10210801 &: 7506890

NB: to recognize and treat severe illness, see p 175. The above carry extra weight in the immunocompromised (eg neonates; post measles; AIDS; cystic fibrosis; leukaemia; chemotherapy/malignancy; absent spleen; B/T cell dysfunction); congenital anomaly or foreign body, eg an indwelling IV line (bacteria may be unusual and of low virulence).

2001 medline:11133897

Fever (Meningitis, p 258; pneumonia, p 278; UTI, p 188). Fever is the major acute presentation in paediatrics, eg from viruses, otitis media, or pharyngitis (p 556), and prognosis is good. Here, fever may be good (aids neutrophil migration; ↑ bioactivity of cytokines, increased production and activity of interferon and increased T-cell proliferation).

2001 medline:11133897

The challenge is to identify serious bacterial infection (see above & below).

Bacteraemia This occurs in ~4% of febrile children, sometimes without an obvious focus of infection; this is made more likely if the child is between 3 and 24 months of age with fevers of $\geq 40^{\circ}\text{C}$, and a WCC $>15 \times 10^9/\text{L}$ (but using these criteria alone, 50% of those with bacteraemia will be missed). ESR $>30\text{mm/h}$ and ↑ C-reactive protein also indicate bacterial infection. Examples of organisms: *Strep pneumoniae*, *N meningitidis*. If blind antibiotics are started (eg shocked), they must cover these organisms (p 258 & p 260). *Cefotaxime*: 50mg/kg/8–12h IV over 20min or *ceftriaxone* 20–45mg/kg once daily IM; use IV route over 30min for top dose (50–80mg/kg/day; max 4g daily); 20–50mg/kg/day IV over 1h if <7 days old).

Finding a focus of infection entails a good history and examination, CXR ± LP (if the child is ill, see above). If ill and no focus of infection is found, it is often appropriate to start blind antibiotics.

Encephalitis (Rarer than septicaemia) Signs: consciousness ↓, odd behaviour, vomiting ± fits. medline:11206692 Infective signs: $T^{\circ} \uparrow$ ± meningism. Consider: herpes simplex (aciclovir, p 256), zoster, mumps, rubella, influenza, measles, rickettsia, toxoplasmosis (OHCM p 576), TB, mycoplasma, Murray Valley encephalitis (Australia), dengue, and rabies. Do CSF PCR; test stool, urine, and blood. 2001 medline:11360206 &: 11247265 &: 11225378 &: 11224846
Noninfective encephalopathies: kernicterus (p 236), hepatic failure (eg Reye's syndrome), lead poisoning, lupus.

medline:11221157

- **UTIs** often present as fever ± D&V: get a 'clean catch' if you can, p 188.
- **Consider malaria** if a malarious area is visited (even for a 'stop-over'). Don't wait to see how the illness unfolds: do a thick blood film at once, and enlist expert help (assume chloroquine resistance, OHCM p 564).
- **Prolonged fevers**—consider: endocarditis; Still's (p 758); malignancy.

Fluid regimens APLS

If tolerated, always use *oral* rehydration. Rehidrat[®] and Dioralyte[®] come in sachets, and contain glucose, Na⁺ and K⁺. Show mother how to make it up (water is the vital ingredient!). If breast feeding, continue.

Daily iv water, Na, and K (mmol/kg/day) MAINTENANCE needs

Age	Weight	Water (mL/kg/day)	Na ⁺	K ⁺
<0.5yr	<5kg	150	3	3
0.5–1yr	5–10kg	120	2.5	2.5
1–3yr	10–15kg	100	2.5	2.5
3–5yr	15–20kg	80	2	2
>5yr	>20kg	45–75	1.5–2	1.5–2

Use dextrose saline for these needs (0.18% NaCl + 4% dextrose; few calories, but prevents ketosis). Pre-existing deficits and continuing loss must also be made good. Reliable input-output fluid balance charts are essential.

Calculating pre-existing deficit mL \approx % dehydration \times weight (kg) \times 10; give eg as 0.45% saline over 24h (eg 750mL for a 10kg child who is 7.5% dehydrated). Add in K⁺ (20mmol/500mL) once the child has passed urine.

Estimating dehydration *Mild dehydration*: Decreased urine output.

5% dehydration: Dry mucous membranes; decreased urine output.

10% dehydration: The above + sunken fontanelle (but if crying, pressure \uparrow), eyeball pressure \downarrow (hard to assess if young); pulse \uparrow ; hoarse cry; skin turgor \downarrow .

>10%: \uparrow severity of the above, with: shock; drowsiness; hypotension. If a recent weight is known, this is useful in quantifying dehydration.

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IV fluids for the first 24h in MILD dehydration If the oral route is impossible, give maintenance needs (above), and start oral fluids when possible.

IV fluids for the first 24h in 5–10% dehydration • Give maintenance water requirement (above) + the deficit. Keep the rate $<25\text{mL/kg/h}$.

- Measure and replace ongoing losses (eg from the bowel).
- Monitor u&e on admission, and at 2, 12, and 24h. Also do pcv.

►► IV fluid replacement in the first 24h in >10% dehydration

- If iv access fails, use the intraosseous route (p 308) and get help to retry ivi.
- 0.9% saline (or plasma, if desperate) at 25mL/kg/h ivi, while calculations are performed. Continuously monitor pulse, BP, ECG.
- Continue until BP rises, pulses are felt, and urine flows (catheterize).
- Then give the daily requirement and the fluid deficit as above, making good continuing loss with 0.18% or 0.45% saline depending on type of dehydration.
- Measure plasma and urine creatinine and osmolality (p 280), and plasma bicarbonate. Metabolic acidosis usually corrects itself.

Guidelines for success: Above all be simple. Complex regimens cause errors.

- 1 Stay at the bedside; use clinical state + biochemistry to adapt ivi.
- 2 Beware sudden changes in Na⁺ (\uparrow or \downarrow); may cause seizures.
- 3 Beware hidden loss (oedema, ascites, GI pools).
- 4 Measure u&e and urine electrolytes often.

Hypernatraemic dehydration: (ie greater water loss than salt, eg from wrongly made feeds, or rarely, if breast-fed). It causes intracellular dehydration (\pm fits, CNS thrombosis/haemorrhages on MRI). medline.10836595
Treatment: rehydrate slowly with 0.45% saline (replace deficit over 48h, lowering Na⁺ by $<12\text{mmol/L/24h}$, and giving only 60% of maintenance volume) to avoid CNS oedema (p 256). The chief danger is too rapid rehydration. Hyperglycaemia is common, but self-correcting. medline.10634988 & 10457791

Water balance in the preterm infant Water comprises 50–70% of weight gain (eg of 15g/day) in preterm babies.

Insensible water loss (IWL) falls with increasing body weight, gestational age, and postnatal age; it increases with $\uparrow T^{\circ}$ (ambient and body) and low humidity. In a single-walled, thermoneutral incubator with a humidity of 50–80% $IWL \approx 30\text{--}60\text{mL/kg/day}$. This may double in infants on phototherapy under a radiant heater.

Faecal water loss $\approx 5\text{--}10\text{mL/kg/day}$ (except during diarrhoea).

Urine loss: If $\sim 90\text{mL/kg/day}$, there is no excessive renal stress.

An intake of 180mL/kg/day (range $150\text{--}200\text{mL}$) of human or formula milk meets the water needs of very low birth weight infants (VLBW; $<1500\text{g}$) under normal circumstances. In infants with heart failure water restriction is necessary (eg 130mL/kg/day).

►►Poisoning: general management

- Find the name and quantity of the poison ingested. The number of tablets dispensed is often recorded on the bottle. The dispenser may also have a record and be able to name the tablets; or try MIMS/BNF. www.emims.net
- When was the poison ingested? Has he or she vomited since? If the child is *comatose*, enlist expert help. Proceed thus:
- Place in the *semi-prone recovery position*. Note the pupil size.
- Have suction, O_2 , a laryngoscope and endotracheal tube to hand.
- Do ward test for *blood glucose*. Treat hypoglycaemia (p 262).
- If there is respiratory failure (do blood gases), summon expert help. Intubate and ventilate. Treat shock (eg with plasma). Note: the mean systolic BP in mmHg (± 1 standard deviation) is 85 (± 15) at birth, 95 (± 15) at 1yr, 100 (± 10) at 4yrs and 110 (± 8) at 10yrs.
- Record level of *consciousness* (coma scale p 680; p 257 if <4yrs). Record *time*.
- If narcotic poisoning is possible, give *naloxone* 10 μ g/kg iv, then 100 μ g/kg every few mins if no response (sc or im all right, but slower action).
- Send blood and urine for a *drug screen*—and blood alcohol if indicated (give 10% dextrose iv: the danger is hypoglycaemia).
- Do a gastric lavage (after intubating the trachea) if ingestion has been recent, or if salicylates suspected. Have the child held with his head lower than his trunk. The length of orogastric tube required is estimated by the length from the mouth to the ear lobe and thence to the xiphisternum. Pass the tube and keep the initial aspirate for analysis. Use gravity to drain 50mL of 1.4% sodium bicarbonate solution into the stomach. Aspirate after a few mins and repeat eg 10 times. Do not use lavage if petroleum products or corrosives have been ingested.
- Monitor TPR, BP, urine output, and blood glucose frequently.

The alert child It is common to induce vomiting by ipecacuanha 10–15mL mixed with 200 mL juice, but this is of uncertain value. medline:10763766
However, activated charcoal (p 254) does reduce absorption.

Specific poisons Get *poisons unit* help. For aspirin & paracetamol, see p 254.

- **Atropine poisoning** (eg from deadly nightshade) causes dilated pupils, dry skin and mouth, tachycardia, excitement \pm psychosis. medline:2330633
With Lomotil® (atropine and diphenoxylate) the pupils may be constricted.
 - **Iron poisoning**: get help. Expect GI symptoms if <20mg/kg elemental iron is ingested (a 200mg $FeSO_4$ tablet \approx 60mg iron). Systemic symptoms, including respiratory distress, occur if >60mg/kg ingested. medline:10664227 &: 8775917
After gastric emptying, give 60mL milk. If >50mg/kg ingested, do iron levels (serum) urgently. If >90 μ mol/L, give desferrioxamine 15mg/kg/h iv; max 80mg/kg/24h. Leave 5g in 50mL water in the stomach after gastric lavage. Beware hypotension. Whole bowel irrigation is tried. medline:10883250
 - **Tricyclic signs**: seizures, coma, cyanosis, arrhythmias (eg if >10mg/kg of amitriptyline is taken); death. Lowest toxic dose: 6.7mg/kg. Monitor ECG (tachycardia + wide QRS). Bretylium may be needed. medline:10696919 &:10452441
 - **Paraffin or petrol (kerosene, gasoline)**: ► Avoid gastric lavage and emetics. Inhalation may cause lung collapse and consolidation. Do CXR. Lung complications may be delayed 12h. Leaded petrol abuse causes encephalopathy, cerebellar and corticospinal signs, dementia, and psychosis. medline:9154447
 - **Toxic alcohols/ethylene glycol**: ?Try fomepizole. medline~11172179 &: 11327228
 - **Sulfonylureas**: ?Try octreotide. medline:10918104 Other antidotes: *OHCM* p 828.
- Contact a National Poisons Information Service (eg 0207 635 9191 UK).

Principles of management

- Support homeostasis by supporting respiration; protect the airway; maintain BP; correct hypoglycaemia; monitor urine output.
- Remove the poison—eg gastric lavage.
- Stop further absorption from the gut—eg charcoal.
- Get help in determining if a specific antidote is available, or if a specific treatment regimen is indicated—eg in aspirin, paracetamol, iron poisoning, and ingestion of corrosives. (The latter requires especial care of the airway, and gastric lavage is contraindicated—see above.)

Salicylate poisoning

Most children can tolerate ingesting 100mg/kg aspirin, and if there is no doubt that a smaller quantity has been ingested and the patient is well, he need not be admitted. Those who have ingested more than this may show deep respiratory movements, causing a respiratory alkalosis, but this is an early sign, before the dangerous metabolic acidosis sets in.

If the child is unconscious, the first step is to place him in the semi-prone recovery position and do a ward test for blood glucose so any hypoglycaemia can be corrected. Protect the airway with an endotracheal tube and perform a gastric lavage (p 252), even if 24h have elapsed since ingestion. Do an urgent plasma salicylate level with blood gases, u&e, and bicarbonate. Blood samples taken within 6h of ingestion may be misleadingly low.

Serious poisoning is indicated by levels $>2.5\text{mmol/L}$ —consider alkaline diuresis (under expert guidance in ITU, as there is significant morbidity) or activated charcoal, which is a much safer means of enhancing the elimination of salicylates. NB: charcoal tastes gritty—and patients may be vomiting after salicylate poisoning, so consider giving by wide-bore nasogastric tube, providing the airway is adequately protected by a cuffed endotracheal tube. A good regimen is 50g (or 1mg/kg body weight) of activated charcoal on admission, or after gastric lavage, then 25g/2–4h, until recovery or until plasma drug concentrations have fallen to safe levels. The dose of charcoal is more related to the quantity of poison ingested than to the size of the child, although the quantity of fluid with which it is mixed may need to be reduced. Charcoal is *not* contraindicated if the child is pregnant. Adding an emetic (ipecacuanha) to this regimen is of no benefit.

An alternative is urinary alkalinization in ITU, eg with small boluses of sodium bicarbonate 1.2% iv; do catheter urine pH every 15min—aim for ~ 8 . If none of these measures is practicable, try haemodialysis or charcoal haemoperfusion (esp. if plasma salicylate is $\uparrow\uparrow$, eg $>5.2\text{mmol/L}$). Seek expert help.

Paracetamol (acetaminophen) poisoning

This is rare in infants as a 100mL bottle of paediatric paracetamol mixture typically contains only 2.4g of paracetamol. Serious hepatotoxicity is likely to occur if $\geq 150\text{mg/kg}$ is ingested. The initial features are nausea and pallor. Jaundice and an enlarged, tender liver occur after 48h.

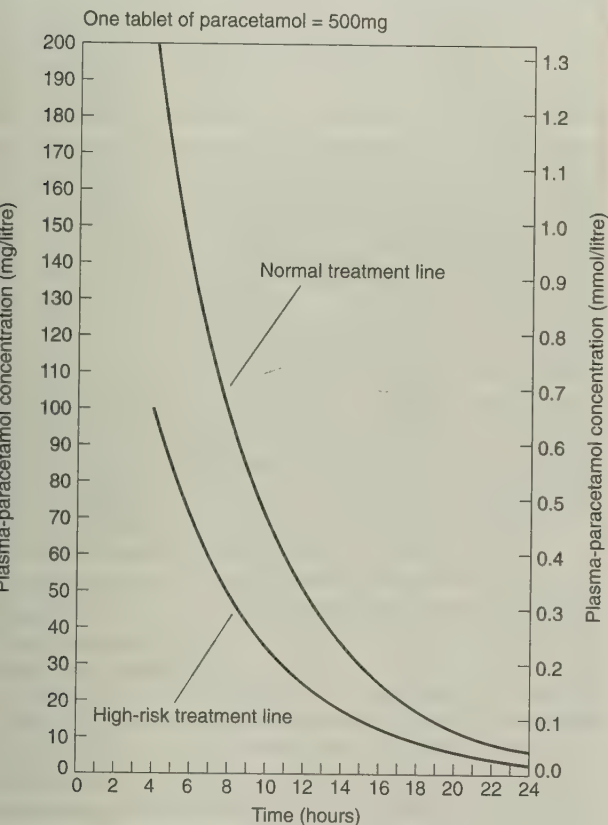
Other features: Hypotension, arrhythmias, excitement, delirium, coma.

Management:

- Empty the stomach if within 2h of ingestion.
- If ingested paracetamol is 140–200mg/kg and ipecac-induced emesis occurs within 1h, management may be safely done at home. medline:10866329 Patients ingesting $>200\text{mg/kg}$ or an unknown amount should be referred for a serum acetaminophen concentration.
- Give N-acetylcysteine if the plasma paracetamol level is above the line in the graph opposite. (Take sample $\geq 4\text{h}$ after ingestion.) The initial dose is 150mg/kg in up to 200mL (depending on the size of the child) of 5% dextrose infused over 30mins, followed by 50mg/kg ivi over the next 4h, and 100mg/kg ivi over the next 16h. This is very effective in preventing liver damage, but only if given within $\sim 8\text{h}$ of ingestion.

Late presentations: Seek expert advice. Check often for hypoglycaemia.

Graph for use in deciding who should receive *N*-acetylcysteine.
 One tablet of paracetamol (acetaminophen) = 500mg; 150mg = 1mmol.



Patients whose plasma paracetamol concentrations are above the 'normal treatment line' should be treated with acetylcysteine by ivi (or, provided the overdose has been taken within 10–12h, with methionine by mouth). Patients who are on enzyme-inducers (eg carbamazepine, phenobarbital, phenytoin, rifampicin, and alcohol) or who are malnourished (eg anorexia, alcoholism, HIV +ve) should be treated if their plasma paracetamol level is above the 'high-risk treatment line'.

We thank Dr Alun Hutchings for permission to reproduce this graph.

►► Rising intracranial pressure^{APL}

Causes Meningoencephalitis; head injury; subdural/extra-dural bleeds (eg abuse); hypoxia (eg near-drowning); ketoacidosis; Reye's syndrome (p 756) may cause sudden icp rises. The rise with brain tumours is slower.

Presentation Listless; irritable; drowsy; headache; diplopia; vomiting; tense fontanelle; ↓level of responsiveness—assess by Children's Coma Scale if <4yrs (see opposite), or by Glasgow Coma Scale if >4yrs (p 680); pupil changes (ipsilateral dilatation); rising BP and falling pulse (Cushing reflex); intermittent respiration. Later, ie chronic: papilloedema and hydrocephalus.

Management The aim is to prevent secondary damage. Help venous drainage by keeping head in the midline, elevated at ~20°. Give O₂. Fan/sponge (tepid water) if T° >40°C. Check for hypoglycaemia. Control any seizures (p 268).

- Intubate and hyperventilate: aim for P_aCO₂ of ~3.68kPa if Coma Scale is <8.
- Give mannitol 20% (check it is crystal-free), 7mL/kg iv over 30min if the child is deteriorating—but discuss this with the neurosurgeon first.
- Dexamethasone 0.1mg/kg iv stat, then 0.05–0.1mg/kg/6h iv.
- Fluid restriction and diuresis, but avoid hypovolaemia (keep Na⁺ 145–150mmol/L, osmolality to 300–310, and cvp to 2–5cmH₂O).
- Measure pulse and BP continuously (check that cuff fits well).
- Remember the possibility of non-accidental injury.

Herpes simplex encephalitis This is the one form of encephalitis discussed in detail as it is treatable. ► *Think of the diagnosis in any febrile child with seizures or CNS (esp temporal lobe) signs ± ↓consciousness.* Patients often present with subtle nonspecific signs. Nasolabial herpes is often absent. CNS deficits may be mild or gross (eg hemiparesis). Focal or generalized fits occur.

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Tests such as CT and CSF examination are often non-specific (unless polymerase chain reaction is available). MRI is better. EEG may show focal changes and periodic complexes. Herpes simplex antibodies are produced too late to guide management. Brain biopsy is now not used. **Treatment:** It is wise to start treatment with aciclovir as soon as the diagnosis is suspected. Give 10mg/kg/8h by iv over 1h (250–500mg/m²) eg for 2 weeks—20mg/kg/8h in neonates. *medline:11269641* Monitor U&E & urine output; adjust dose according to Datasheet. **Mortality:** ~7%. 60% survive without CNS sequelae. There may be Klüver–Bucy syndrome (hypersexuality + episodic rage + visual agnosia).⁵⁴

Brain tumours 2/3 are in the posterior fossa. Key features in their diagnosis are a raised icp with focal signs (below), ± false localizing signs (eg VI nerve palsy, due to its long intracranial course). Space-time clustering suggests a viral cause of astrocytomas and ependymoma. *Br J Cancer 2002 86 1070*

Medulloblastoma: Radiosensitive midline cerebellar tumour (inferior vermis) causing truncal ataxia ± falls. ♂/♀≈4:1 Seeding is along CSF pathways.

Brainstem astrocytoma: Cranial nerve palsies; pyramidal tract signs (eg hemiparesis); cerebellar ataxia; signs of icp↑ are rare.

Midbrain and third ventricle tumours may be astrocytomas, pinealomas or colloid cysts (cause posture-dependent drowsiness). Signs: behaviour change (early); pyramidal tract and cerebellar signs; upward gaze defect.

Suprasellar gliomas: Visual field defects; optic atrophy; pituitary disorders (growth arrest, hypothyroidism, delayed puberty); diabetes insipidus (DI). Cranial DI is caused by ADH↓, so that there is polyuria and low urine osmolality (always <800mosmol/L) despite dehydration.

Cerebral hemispheres: Usually gliomas. Meningiomas are rare. Fits may be the presenting sign. Signs depend on the lobe involved (*OHCM* p 373).

Tests: EEG, skull x-ray, CT scans ± magnetic resonance imaging.

Treatment options: Excision, CSF shunting, radio- and chemotherapy.

- Headaches of increasing frequency or severity.
- Age <6yrs.
- Headache unrelieved by paracetamol.
- Irritable; loss of interest/skills.
- Slowing of physical or cognitive development.
- Head circumference above the 97th centile, or greatly out of line.

Children's coma scale¹ (use if <4yrs)^{APLS}

The is an objective scale to record the level of coma, and to help answer questions such as: 'Is she getting better or worse; what is the prognosis?'

Best motor response (6 grades)

- 6 *Carrying out request ('obeying command'):* The child moves spontaneously, or to your request.
- 5 *Localizing response to pain:* Put gentle pressure on the patient's finger nail bed with a pencil then try supraorbital and sternal pressure: purposeful movements towards changing painful stimuli is a 'localizing' response.
- 4 *Withdraws to pain:* Pulls limb away from painful stimulus.
- 3 *Flexor response to pain:* Pressure on the nail bed causes abnormal flexion of limbs—decorticate posture.
- 2 *Extensor posturing to pain:* The stimulus causes limb extension (adduction, internal rotation of shoulder, forearm pronation; decerebrate posture).
- 1 *No response to pain.* Score best response of any limb.

Best verbal response (5 grades) If intubated *can the patient grimace?*

- | | |
|---------------------------------------------------------------------------|---------------|
| 5 Orientated: Smiles, is orientated to sounds, fixes and follows objects. | |
| Crying | Interaction |
| 4 Consolable | Inappropriate |
| 3 Inconsistently consolable | Moaning |
| 2 Inconsolable | Irritable |
| 1 No response | No response |
- Score the highest out of either columns.

Eye opening (4 grades)

- 4 Spontaneous eye opening.
- 3 Eye opening in response to speech: Any speech, or shout, not necessarily request to open eyes.
- 2 Eye opening to response to pain: Pain to limbs as above.
- 1 No eye opening.

An overall score is made by summing the score in the 3 areas assessed. Eg: no response to pain + no verbalization + no eye opening = 3.

- If >4 yrs, see *Glasgow Coma Scale*, p 680; scores may be hard to interpret.¹ As a rule of thumb, a score of >7–8 means intubation may be needed; 4–8 means intermediate prognosis; 3 means a bad prognosis.

►►MeningitisAPLS

Suspect this in any ill baby or child. The only signs may be unusual crying, poor feeding, or vomiting. Pay great attention to the fontanelle: in GI vomiting it becomes sunken; with meningitis it may be tense. ►Get expert help from your senior and a microbiologist; if confirmed, notify your local consultant in Communicable Disease Control (ccdc). If there is any hint of meningococcal disease, give penicillin at once (dose below). On lumbar puncture, if csf is cloudy, start a broad-spectrum iv agent (eg ceftriaxone below) at once, while awaiting microscopy and further advice.

Meningeal signs: Stiff neck (unable to kiss knees), often absent if <18 months old; photophobia; Kernig's sign (resistance to extending knee, with hip flexed); Brudzinski's sign (hips flex on bending head forward); opisthotonos.

ICP↑: Irritable; high-pitched cry; drowsy; vomiting; fontanelle tense (late sign).

Septic signs: Fever; arthritis; odd behaviour; rashes—any, do not expect petechiae with meningococcus; cyanosis; DIC; pulse↑; BP↓; tachypnoea; wcc↑.

Other causes of meningism: Tonsillitis, pneumonia, subarachnoid bleed.

Lumbar puncture: Do this at once unless: focal signs; DIC; purpura (top priority is penicillin 50mg/kg/4h iv); or if brain herniation is near (abnormal postures or breathing; coma scale <13, p 257; dilated pupils, doll's eye reflexes, BP↑, pulse↓, papilloedema). NB: preliminary CT is incapable of showing that LP would be safe. **Technique:** Learn from an expert; a common error is aiming too low. • Explain everything to the mother. • Ask an experienced nurse to help, by positioning the child fully flexed (knees to chin) on the side of a bed, with his back exactly at right angles with it. • Mark a point just above (cranial to) a line joining the spinous processes between the iliac crests. • Drape and sterilize the area; put on gloves. • Infiltrate 1mL of 1% lidocaine. • Insert LP needle aiming towards umbilicus. • Catch 4 csf drops in each of 3 bottles for: urgent Gram stain, culture, virology, glucose, protein (do blood glucose too). • Request frequent CNS observations. • Report to mother. **Tests:** (after LP): FBC, U&E, culture blood, urine, nose swabs, stool virology. C-reactive protein: is it >20mg/L? CXR. Fluid balance, TPR & BP hourly. csf lactate: is it >3mg/L? (may be a more sensitive means of distinguishing bacterial from aseptic meningitis than the blood/csf glucose ratio).

Treating pyogenic meningitis before the organism is known

- Protect airway and give high flow O₂
- Set up ivi: colloid in 10mL/kg boluses.
- If meningococcaemia likely use antibiotics as detailed on p 260.
- Ceftriaxone 80mg/kg/day ivi over 30min (50mg/kg if <7 days old, over 1h). Alternative: cefotaxime up to 50mg/kg/6h iv over 15min. Benzylpenicillin may also be used—50mg/kg/8h slowly iv if 1 week to 1 month old; 50mg/kg/4h if older. Netilmicin may be used in neonates (2mg/kg/8h slowly iv—monitor plasma levels, U&E, and creatinine, ↓dose as needed; see Data sheet); with ampicillin in case of listeria.

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NB: in general, the intrathecal route is not required for antibiotic therapy.

- If pre-hernia signs, treat for ICP↑ (p 256), eg mannitol 20%, 2.5mL/kg ivi.
- Once the organism is known and the right drug started, find its minimum inhibitory concentration (MIC) to the organism *in vitro*.

Complications Disseminated sepsis, subdural effusion, hydrocephalus, ataxia, paralysis, deafness, mental retardation, epilepsy, brain abscess.

CSF in meningitis

	Pyogenic:	Tubercular:	Aseptic:
Appearance	often turbid	may be fibrin web	usually clear
Predominant cell	polymorphs	mononuclear	mononuclear
Glucose level	eg 1000/mm ³	10–350/mm ³	50–1500/mm ³
Protein	< 2/3 of blood	< 2/3 of blood	> 2/3 of blood
	1–5g/dL	1–5g/dL	< 1.5g/dL

Preventing deaths from meningococcal disease

Because death is so swift, this can seem a hopeless task—as even when we doctors contract meningococcal disease, we simply think we have 'flu, and our families are surprised to find us dead in bed just hours after reporting sick. Nevertheless, good practice (and luck) *can* save lives, and the meningococcus is not always *that* fast.

- ▶ Do not expect meningeal signs always to be to the fore. Septicaemic features predominate in fatal illness.
- ▶ Any rash (or none) will do for the meningococcus. If you wait for purpura, you may be waiting until the disease is untreatable.
- ▶ In primary care, for any acutely unwell child *leave your consulting room door ajar*. Explain that a doctor can be contacted *at any time* if:
 - He seems to be worsening –He seems to be unrousable
 - A rash develops –He starts crying in an odd way
- ▶ Beware fever+lethargy+vomiting, even if no headache or photophobia.
- ▶ Give parenteral penicillin *early*, before admission to hospital.
- ▶ Ensure immunization (the new protein–polysaccharide conjugate vaccine prevents type C but not the commonest type which is B; see p 209).
- ▶ Inform your local CDC (consultant in communicable diseases).
- ▶ Arrange prophylaxis (rifampicin, ciprofloxacin or ceftriaxone) for household contacts and those in contact with secretions.

medline:10541486

See *OHCM* p 737 for heroic, non-standard ways of augmenting management, eg extracorporeal membrane oxygenation; use of terminal fragment of human bactericidal/permeability-increasing protein (rBPI₂₁) to ↓cytokines; heparin with protein C concentrate to reverse coagulopathy; plasmapheresis to remove cytokines, and correct acidosis—and thrombolysis (rTPA) to help limb re-perfusion.

medline:9393331

Meningitis: the organisms

The meningococcus, the pneumococcus, and, in the unvaccinated, *Haemophilus influenzae* are the great killers, but of these it is the meningococcus that is the swiftest and the most deadly. The interval between seeming wellbeing and coma may be counted in hours. If you suspect meningitis, give benzylpenicillin (opposite) before hospital admission (don't worry about ruining the chance of a +ve blood culture later).

Neisseria meningitidis Abrupt onset \pm rash (purpuric or not, eg starting as pink macules over legs). Septicaemia may occur without meningitis (Waterhouse–Friderichsen, *OHCM* p 736), so early LPs may be normal, giving false reassurance. Arthritis, conjunctivitis, myocarditis, and DIC may coexist.

Typical age: Any. **Film:** Gram –ve cocci in pairs (long axes parallel), often within polymorphs. **Drug of choice:** benzylpenicillin (G) 50mg/kg/4h iv. If penicillin-allergic, give cefotaxime (p 258) or chloramphenicol (below). Treat shock with colloid (\pm inotropes). **Prevention:** Rifampicin 10mg/kg/12h po (if <1yr, 5mg/kg/12h) for 48h for close contacts, or ceftriaxone 125mg IM if under 12yrs (250mg IM stat if over 12yrs). Observe closely.

Haemophilus influenzae (Rare if immunized). **Typical age:** <4yrs. **Gram stain:** Gram –ve rods. The lower the CSF glucose, the worse the infection. **Drugs:** Ceftriaxone (p 258) or, where resistance is not a problem, chloramphenicol 12–25mg/kg/6h ivi (neonates: see *BNF*) with ampicillin 30mg/kg/4h iv. Rifampicin (below) may also be needed. If chloramphenicol is used, monitor peak levels; aim for 20–25 μ g/mL; usual doses may be far exceeded to achieve this. (Trough level: <15 μ g/mL.) As soon as you can, switch to po (more effective). Dexamethasone (0.15mg/kg/6h iv for 4d) has been reported by some to reduce post-meningitis deafness.

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Strep pneumoniae (eg via respiratory infections, skull fracture, meningocoele). **Typical age:** Any. **Film:** Gram +ve cocci in pairs. **R_x:** Benzylpenicillin 25–50mg/kg/4h slow iv—or, if resistance likely (eg parts of Europe, USA), vancomycin (dose if >1 month old: 10mg/kg/6h ivi over 1h) with a 3rd-generation cephalosporin (ceftriaxone, p 258) is advised. Monitor U&E.

Escherichia coli This is a cause of meningitis in the neonate (in whom the signs may consist of feeding difficulties, apnoea, fits, and shock). **Drug of choice:** cefotaxime (p 258) or netilmicin 2mg/kg/8h slowly iv (p 238).

Group B haemolytic streptococci eg via mother's vagina (so swab mothers whose infants suddenly fall ill at ~24h-old). Infection may be delayed for a month. **Drug of choice:** benzylpenicillin 25–50mg/kg/8–12h slowly iv.

Perinatal listeriosis (*Listeria monocytogenes*) This presents soon after birth with meningitis and septicaemia (\pm pneumonia). Microabscesses form in many organs (granulomatosis infantiseptica). **Diagnosis:** Culture blood, placenta, amniotic fluid, CSF. **Treatment:** iv ampicillin with gentamicin.

TB can cause CNS infarcts, demyelination (hence cranial nerve lesions) and tuberculomas \pm meningitis (long prodrome: lethargy, malaise, anorexia). Photophobia/neck stiffness are likely to be absent. The first few CSFs may be normal, or show visible fibrin webs, and widely fluctuating cell counts.

Treatment: get help. **Dose example:** Isoniazid 10mg/kg/24h iv with vit. B₆ + rifampicin 10mg/kg/24h (\leq 600mg/day) for 1yr + (for 2 months) pyrazinamide 35mg/kg/day + eg streptomycin 15mg/kg/day IM adjusted to give a peak plasma level <40 μ g/mL and a trough of <3 μ g/mL; alternative ethambutol 15mg/kg/day po if old enough to report visual problems. medline: 10675209

Other bacteria Leptospiral species (canicola); Brucella; Salmonella.

Causes of 'aseptic' meningitis Viruses (eg mumps, echo, herpes, polio), partially treated bacterial infections, cryptococcus (use ink stains).

Giving benzylpenicillin* before hospital admission

- 300mg IM up to 1 year old
- 600mg if 1–10yrs
- 1.2g if >10yrs

► When in doubt, give it: it may be negligent not to do so.

*If penicillin-allergic, cefotaxime may be tried (20–50mg/kg IM) [@11826463]

Diabetes mellitus (DM)—some questions

The new diabetic (type I) *Presentation:* Infection (eg UTI), poor growth, ketoacidosis (rare). *Typical age:* 4–12yrs. *Diagnostic criteria:* OHCM p 282. *Rx:* If nonketotic, iv fluids are rarely needed. Suitable initial dose of insulin: 0.2u/kg sc followed by 0.35–0.5u/kg/12h sc, 30min before breakfast and tea. ~70% of the dose should be long-acting. With help from the pancreas a 'honeymoon' period may follow in which insulin needs fall to 0.1u/kg/day. *Diet:* Ask a paediatric dietitian to help. 1500kcal/m² or 1000kcal plus 100–200 kcal for each year of age (depending on energy used). Aim for 30% of this with each major meal, and 10% as a bedtime snack. Give ~20% of calories as protein, ~50% as unrefined carbohydrate, and ≤30% as fat.

Type II DM Insulin resistance is still rare in children.

medline:11713450

What should the family know before the child leaves the ward?

- Insulin: doses (eg during illness); self-injecting skills (practise on oranges).
- Diet: What is it? Why is it important? Why must meals be regular? What does the carer/parent do if the child is hungry?
- Can blood sugar be monitored accurately? Watch the carer's technique.
- What does the carer do if the blood sugar is not well controlled?
- Does the parent or carer know what 'well controlled' means?
- What happens if he has too much insulin? What will happen as hypoglycaemia evolves? (Weakness, hunger, irritability, faintness, sweating, abdominal pain, vomiting, fits, and coma). What action needs to be taken? Practise inducing hypoglycaemia (glucose <2mmol/L) on the ward, by omitting breakfast after morning insulin. Explain symptoms as they happen, and their amelioration with oral drinks (or Hypostop® oral jelly). If things get out of hand, for *hypoglycaemic coma*, give glucose 0.5 g/kg as a 25% ivi (or via rectal tube if no iv access, with glucagon 0.5–1mg im or slowly iv). Then give 10% dextrose, 5mL/kg/h. Expect a quick return to consciousness. If not, recheck glucose; if low, give iv dexamethasone (p 256); if normal, ask yourself if there is a post-ictal state after a hypoglycaemic fit: don't give more glucose as this can promote cerebral oedema.
- What should happen if the child misses a meal, or is sick afterwards? What happens to insulin requirements during 'flu and other illnesses?
- Who does mother contact in emergency? Give written advice.
- Is the GP informed of discharge and follow-up arrangements?
- Encourage membership of a *Diabetic Association* (uk tel: 0207 323 1531).

'What are the aims of routine follow-up in the diabetic clinic?'

- To achieve normoglycaemia through education and motivation.
- To prevent complications. So check growth and fundi (dilate pupils; NB: retinopathy usually takes 10yrs to develop). Blood: glucose, HbA1c.
- If normoglycaemia is unachievable, choose the best compromise with the child's way of life and strict glucose control. Insulin-storing pen-shaped injectors allow great flexibility in the timing and dose of insulin, delivering a variable dose (2u/push) at the push of a plunger, and obviates the need for carrying syringes and drawing up insulin at inconvenient times (eg during a party). Get help from your local diabetic nurse.

Genes & environment Diabetes used to be a disease of the over-5s, but in the UK, incidence is rising in infancy from ~1/10,000 in 1985–90 to ~1/6/10,000 in 1995. Islet cell antibodies are found in HLA-B8 (but not HLA-B15) individuals. If one child in a family has DM, risk to siblings is >5%. * Mean duration of symptoms pre-diagnosis is 30 days. Feeding with cows' milk, and infant exposure to enteroviruses, may ↑ incidence. 10% have some degree of coeliac disease, so screening is wise. medline:11808892. We note with interest that twin studies show that, although at least 4 genes are thought to be important—and one (6q) partly determines islet sensitivity to damage, most of the susceptibility to diabetes is environmentally acquired, and patients with type I diabetes have antibodies to cows' milk albumin which react with β-cell (the insulin-producing cell) surface proteins. We also know that mumps, rubella, and coxsackie viruses (± CMV) have potential to injure β-cells—but their exact rôle is unclear.

Childhood diabetic ketoacidosis (DKA)¹

The Patient Listlessness; confusion, vomiting; polyuria; polydipsia; weight loss; abdominal pain. **Look for:** Dehydration; deep and rapid (Kussmaul) respirations; ketotic (fruity-smelling) breath; shock; drowsiness; coma.

Diagnosis Usually straightforward, based on the combination of hyperglycaemia (may be mild as capillary blood glucose may be falsely low if the patient is vasoconstricted), acidosis and ketones in urine. Where there is a severe metabolic acidosis without hyperglycaemia (or other obvious causes of acidosis, eg renal failure); consider these other causes: lactic acidosis (including glycogen storage disease Type I), alcoholic ketoacidosis, salicylate overdose, other inborn errors of metabolism (propionic acidemia, methylmalonic acidemia) and septicaemia (eg Gram -ve).

Resuscitation Do coma score (p 680; p 257 if <4yrs). True coma is rare (<10%) in DKA: exclude other causes of coma and remember DKA may have been precipitated secondarily. Very rarely cerebral oedema may occur before iv treatment).¹ If consciousness ↓, nurse semi-prone, to prevent aspiration. Take the following action if shocked, consciousness ↓, coma, or vomiting:¹

- Airway: oro-pharyngeal airway ± nasogastric tube.
- Breathing: 100% O₂.
- Circulation: up to 3 10mL/kg boluses of 0.9% saline iv (or 4.5% albumin).
- Take to ITU if shocked, very acidotic, very young, or ward staff over-busy etc.
- Weigh; FBC; U&E; glucose; Ca²⁺; PO₄³⁻; blood gas; ECG monitoring.
- Use clinical signs to assess dehydration (capillary refill time >3sec ≈ >10% dehydration). Calculate the volume of fluid to be replaced (fluid requirement): ie maintenance fluid (basal 24 hour requirement—eg 100mL/kg/day) + the water deficit. It should be given at a constant steady rate over the first 24 hours. Ongoing losses (eg vomiting) should be replaced if excessive and balance remains negative. Use a maximum of 10% dehydration for the initial calculations (10% dehydrated = water deficit of 100mL/kg).
- If child is <5% dehydrated and tolerating oral fluids; it may be appropriate to start with oral fluids and sc insulin rather than iv regime below.
- Aim to complete rehydration over 24h (longer if very young or plasma glucose ↑↑—rapid infusions of large volumes may risk cerebral oedema).*
- 0.9% saline is used at the start of iv therapy until blood glucose falls to around 12 mmol/L: then change to 0.45% saline + dextrose 5%.²
- Insulin (eg Actrapid®). If >5yrs: 0.1u/kg stat iv, then 0.1u/kg/h ivi by pump in 0.9% saline. If no pump, give 0.25u/kg im stat, then 0.1u/kg/h im. Halve dose to 0.05u/kg/h if glucose falls faster than 5mmol/L/h. If <5yrs old, consider using a lower dose (0.05u/kg/h)—but this may be insufficient to switch off ketogenesis. Don't switch off the insulin ivi if the blood glucose falls; add more dextrose to the ivi; insulin is needed to reverse ketosis.
- Add ivi K⁺, 20mmol for every 500mL,[†] as guided by U&E. The danger is hypokalaemia; aim for plasma K⁺ of 4–5mmol/L.

There is no evidence favouring bicarbonate in DKA. If acidosis persists, consider giving further colloid, or ↑dose of insulin (more glucose may be needed in the iv fluids). Ask yourself: is this child septic?

Ongoing monitoring • Hourly blood glucose • CNS status ≥half-hourly.

• Nurses must tell you of headache or behaviour change promptly as these may indicate cerebral oedema • Hourly fluid input/output • U&E + blood gases (for acid/base balance) 2h after starting ivi, then 4-hourly. • Weigh twice daily • Monitor ECG for T-wave changes • CXR • Infection screen.

When drinking well, tolerating food (urine ketones may still be positive); start subcutaneous insulin—then stop intravenous insulin one hour later.

Neurological deterioration warning signs in DKA

If warning signs (headache, irritability, falling consciousness, rising ICP):

- ▶ Call your senior
- ▶ Exclude hypoglycaemia
- ▶ Give mannitol 0.5g/kg IV immediately
- ▶ Restrict IV fluids by two-thirds
- ▶ Move to ITU and perform CT scan at an opportune moment
- ▶ Treat any possible sepsis (see below)

If there is no improvement—re-evaluate fluid balance and IV therapy. If acidosis continues, further colloid or larger insulin doses may be required.

Pitfalls in diabetic ketoacidosis

- WCC ↑ (even a leukaemoid reaction) may occur without any infection.
- Infection: often patients are apyrexial. Perform MSU, blood cultures, and CXR. Start broad spectrum antibiotics (p 258) early if infection suspected.
- Creatinine: some assays for creatinine crossreact with ketone bodies, so plasma creatinine may not reflect true renal function.
- Hyponatraemia may occur from osmotic effects of glucose; if $<120\text{mmol/L}$, search for other causes, eg triglycerides ↑. Hyponatraemia $>150\text{mmol/L}$ may be treated with 0.45% saline to start with (0.9% saline thereafter).
- Ketonuria does not equate with ketoacidosis. Normal individuals may have ketonuria after an overnight fast. Not all ketones are due to diabetes—consider alcohol, if glucose normal. Test plasma with Ketostix® or Acetest® to demonstrate ketonaemia.
- Acidosis without gross elevation of glucose may occur, but consider poisoning, eg with aspirin).
- Serum amylase is often raised (up to 10-fold), and nonspecific abdominal pain is common even in the absence of pancreatitis.
- Cerebral oedema is a constant threat, and is almost exclusively a condition of childhood. Pathophysiology is incompletely understood: it usually occurs between 4–12h from the start of treatment, but it may be present at onset of DKA or up to 24h afterwards, presenting as a sudden neurological deterioration after an initial gradual general improvement.

*Because thrombotic complications are rare, heparin is not usually required and is not recommended as part of standard therapy, but see [10332664] and www.brighton-health-care.nhs.uk/diabetes/newpage17.htm re: thrombotic thrombocytopenia [8123958].

†Note that pooling of KCl and insulin may cause uneven delivery in some IV containers, so repeated mixing may be needed. [6755202]

1 Guidelines for the management of DKA in children & adolescents; Diabetes UK 2002 & D Dunger 1999 Ketoacidosis; Practical Algorithms in Ped Endo www.diabetes.org.uk/dka_paed/dkapaed.pdf

2 0.45% saline is preferable to 0.18% saline, as there is some evidence that falling plasma sodium concentrations may be associated with cerebral oedema (see reference 1 above).

We thank Dr Huw Thomas for drafting this page.

Epilepsy and febrile convulsions

EPILEPSY is a tendency to intermittent abnormal electrical brain activity. Classification depends on whether signs are referable to one part of a hemisphere (partial epilepsy) or not (generalized), and on whether consciousness is affected (complex) or not (simple). The 4 kinds of *generalized epilepsy*:

- **Tonic/clonic (grand mal):** Limbs stiffen (the tonic phase) and then jerk forcefully (clonic phase), with loss of consciousness.
- **Absences.** Brief (eg 10sec) pauses ('he stops in midsentence, and carries on where he left off'); eyes may roll up; he/she is *unaware of the attack*.
- **Infantile spasms:** Eg at 5 months. Jerks forwards with arms flexed, and hands extended ('Salaam attack'). Repeated every 2–3sec, up to 50 times. Here the EEG is characteristic. Treat with valproate or vigabatrin.
- **Myoclonic fits:** 1–7-year-olds; eg 'thrown' suddenly to the ground.

Partial epilepsy Signs are referable to part of one hemisphere. Complex phenomena: (temporal lobe fits) consciousness↓; automatisms (lip smacking, rubbing face, running); fits of pure pleasure. medline:7439135 & 3339342

Causes Often none is found. Infection (eg meningitis); $U\&E\uparrow$, glucose↓, $Ca^{2+}\downarrow$, $Mg^{2+}\downarrow$; $Na^+\uparrow\downarrow$; toxins; trauma; metabolic defects; tuberous sclerosis; CNS tumour (<2%) or malformation; flickering lights, eg TV; exercise. medline:9070589

Differential diagnosis: Arrhythmias, migraine, narcolepsy, night terrors, faints (reflex anoxic seizures, p 267), tics, Münchausen's (eg by proxy, *OHCM* p 730).

Tests Expert EEG; CT eg in infantile spasms, unusual seizures, CNS signs, or partial or intractable epilepsy. MRI is more sensitive but less available and may require anaesthesia or sedation—but there is no radiation.

A FEBRILE CONVULSION is diagnosed if the following occur together:

- A tonic/clonic, symmetrical generalized seizure with no focal features
- Occurring as the temperature rises rapidly in a febrile illness
- In a normally-developing child between 6 months and 5yrs of age
- With no signs of CNS infection or previous history of epilepsy
- When there are fewer than 3 seizures, each lasting <5 min.

Differential: Meningoencephalitis, CNS lesion, trauma, glucose↓, $Ca^{2+}\downarrow$; $Mg^{2+}\downarrow$.

Lifetime prevalence: ~3% of children have at least one febrile convulsion.

Examination: A complete search to find any possible infection.

Tests. FBC, MSU, CXR, ENT swabs, LP (always if aged <1 year). Get help in deciding if an LP is needed. If it is not, review this decision in ~2h. If LP is indicated, be sure to exclude signs of $ICP\uparrow$ (see p 258).

Management: Lie prone; diazepam (p 268). Tepid sponging if hot; 12mg/kg/6h paracetamol syrup, 120mg/5mL. **Parental education:** Allay fear (eg a child is *not* dying during a fit). Febrile convulsions don't usually mean risk of epilepsy. For the 30% who have recurrences, try teaching mother to use rectal diazepam, eg with a 5mg tube (Stesolid®) if 1–3yrs*, or a 10mg tube if older. This dose may also be used at home *during* seizures before calling for help.

Further prevention: Prophylactic anticonvulsants are almost never needed.

Follow-up: Explain that fever with future pertussis and MMR vaccination should prompt oral paracetamol (p 214)—with rectal diazepam to hand.

Prognosis: If no focal signs, and the seizure lasts <30min, and is single only, 1% develop epilepsy. <1% have all these adverse signs: their risk of epilepsy is much higher (50%, eg if coupled with pre-existing CNS abnormality).

Treatment and prevention of epilepsy See p 268 and p 269.

* If <3yrs insert PR half way to mark on the nozzle. ~2.5mL is expellable (don't worry about the bit left behind: this is allowed for in the dose formulation) NB: CP **diazepam Rectube®** dose differs: its licence is for a 0.5mg/kg **single** dose (adults and children >10kg—a typical weight for a 1yr-old), **which shouldn't be repeated in <12h** (there are very few reports of respiratory problems at this dose: all survived).

Specific types of seizure

Reflex anoxic seizures Paroxysmal, self-limited brief (eg 15sec) asystole triggered by pain, fear, (eg at venepuncture) or anxiety, or an overwhelming confrontation with reality, perhaps in the form of a bath which is unexpectedly hot or cold. During this time the child is deathly pale— \pm hypotonia, rigidity, upward eye deviation, clonic movements, and urinary incontinence. Typical age: 6 months to 2 years (but may be much older).

Prevalence: Maybe 0.8% of preschool children.² **Differential** The most often confusing condition is epilepsy—which is frequently misdiagnosed, because the trigger aspect to the history is ignored or unwitnessed. When in difficulty, refer to a specialist for vagal excitation tests, while under continuous EEG and ECG monitoring (ocular compression induces the oculocardiac reflex; do not do this test if there is glaucoma, or known arrhythmia, or if you lack experience: it is uncomfortable for the child). **NB:** tongue-biting is not described in reflex anoxic seizures. **Management:** Drugs are rarely, if ever, needed. Atropine has been tried, to reduce sensitivity to vagal influences. Anticonvulsants are not indicated. Inserting a pacemaker is one option under review. **What to tell parents:** Avoid the term 'seizure', as this is all that is likely to be remembered, however careful your explanation. *Pallid syncopal attack*, or *white breath-holding attacks* are useful synonyms. Emphasize the benign nature of the condition, and that the child usually grows out of it (but it may occur later in life, and in older family members).

medline:8369674

Panayiotopoulos syndrome (eg 6% of those with seizures—peak age: 5ys). It occurs mainly at night, with vomiting and eye deviation with impaired consciousness before the convulsion starts. Many seizures last for 30mins (some may last hours)—but there is no permanent brain damage.

Diagnosis: EEG. **Treatment:** As remission often occurs within 2 years, antiepileptic medication is often not needed. Reassure.

medline:11265947

Rolandic epilepsy (benign epilepsy with centrotemporal spikes, BECTS) 15% of all childhood epilepsy. Infrequent, brief partial fits with unilateral facial or oropharyngeal sensory-motor symptoms, speech arrest \pm hypersalivation. Sulthiame may be used in some units.

medline:11231230 & 11051123

Stepwise treatment of status epilepticus Supportive therapy: •Secure airway; give O₂. Set a clock in motion. •Check T°; if ↑, give rectal diazepam and rectal paracetamol (it may be a febrile convulsion) •Set up 5% dextrose ivi •Do BP, pulse, glucose, Ca²⁺ (± Mg²⁺). ►If hypoglycaemic, give glucose 0.5g/kg iv as a 25% solution, then 5–10mg/kg/min as 10% dextrose ivi.

Seizure control: Proceed to the next step only if fits continue.

- 0min Estimate the weight of the child: $\text{kg} = 2 \times [\text{age in yrs} + 4]$.
Lorazepam 50–100µg/kg iv (large vein). If seizures continue, *paraldehyde* 0.3mL/kg PR in arachis oil is an option: max 10mL PR; avoid contact with plastics/rubber. See also diazepam PR, p 266.
 20min *Phenytoin* 18mg/kg ivi; max rate 1mg/kg/min. Monitor ECG.
 50min *Diazepam* 0.1–0.4mg/kg/h ivi. Get expert help. Transfer to ITU.
 65min Paralyse and ventilate: use *thiopental* (=thiopentone) infusion.

►These times refer to elapsed time on the clock from the first drug, not gaps between each drug. Some authorities recommend starting ventilation earlier, and always be ready to do this to protect the airway.

Tests S_AO₂, ECG monitor, glucose, U&E, Ca²⁺, Mg²⁺, arterial gases, FBC, platelets, ECG. Consider anticonvulsant levels, toxicology screen, blood ammonia, lumbar puncture, culture blood and urine, virology, EEG, MRI CT, carbon monoxide level, lead level, amino acid levels, metabolic screen.

Once the crisis is over, start prophylaxis, eg with sodium valproate or carbamazepine, guided by regimen on p 269. Aim to use one drug only. Increase dose until fits stop, or toxic levels reached. *Out of the context of status, it is not known when to start prophylaxis.* After 2–3 fits is a rule of thumb.

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Carbamazepine *SE* Rash (± exfoliation); platelets↓, agranulocytosis, aplasia (all rare). It induces its own enzymes, so increasing doses may be needed.

Sodium valproate The sugar-free liquid is 200mg/5mL. *SE:* vomiting, appetite↑, drowsiness, thrombocytopenia (do FBC pre-treatment). Rare hepatotoxicity can be fatal (eg if co-existing unsuspected metabolic disorder). Routine monitoring of LFTs is needed, usually only in first 6 months of use.

Ethosuximide The syrup is 250mg/5mL. *SE:* D&V, rashes, erythema multiforme, lupus syndromes, agitation, headache. Indication: absence epilepsy.

Lamotrigine This is increasingly used in absences and intractable epilepsy as an add-on. **When given with valproate** the dose is 0.2mg/kg/24h po for 2 weeks, then 0.5mg/kg daily for 2 weeks, then increasing by up to 0.5mg/kg every week. Usual maintenance dose: 1–5mg/kg/day (higher if combined with non-valproate anticonvulsants—eg 2.5–7mg/kg/12h po).

Vigabatrin (May be 1st choice in infantile spasms and tuberous sclerosis fits.) This blocks GABA transaminase. Consider adding it to regimens if partial seizures are uncontrolled. Vigabatrin starting dose: 40mg/kg/day po (sachets and tabs are 500mg). Max: 100mg/kg/day. Blood levels do not help (but monitor concurrent phenytoin: it may fall by ~20%). *SE* Drowsiness, depression, psychosis, amnesia, diplopia, and field defects (test every year).

Phenytoin Rarely used as dosage is difficult and cognitive deterioration, depression, motivation↓, behavioural disorders, polyneuropathy, acne, coarse facial features, hirsutism, gum hypertrophy, and blood dyscrasias are problems.

Education If fits are few, educate teachers on lifting bans on supervised swimming, and cycling etc. Showers are safer than baths. Emphasise importance of compliance (fits may ↓threshold for further fits: the concept of *kindling*).

Drug	Starting dose mg/kg/24h	Target dose for initial assessment of effect mg/kg/24h	Dose increment		Usually effective dose mg/kg/ 24h	Doses per day	Target trough drug level in plasma mg/L $\mu\text{mol/L}$	
			Size mg/kg/ 24h	Interval in days			mg/L	$\mu\text{mol/L}$
Carbamazepine	5	12.5	2.5	7	10-25	2-3	4-10	17-42
Valproate	10	20	10.0	10	15-40	1-2	Not helpful	
Phenytoin	5	7	1.0	10	5-9	1-2	10-20	40-80
Phenobarbital	4	6	2.0	10	4-9	1	15-40	60-180
Ethosuximide	10	15	5.0	5	15-40	1	40-99	280-700
Clonazepam	0.025	0.05	0.025	7	0.025-0.1	2-3	Not helpful	

When to use which drug


Tonic/clonic fits: first try sodium valproate or carbamazepine.

Absences: first choice, ethosuximide; second, sodium valproate.

Myoclonic or akinetic fits: sodium valproate or benzodiazepines.

Infantile spasms: prednisolone or nitrazepam (0.2-0.6mg/day po).

Partial fits: first try carbamazepine; then sodium valproate.

Stopping anticonvulsants See *OHCM* p 367. The risk of seizure recurrence during the tapering down process is no greater if the tapering period is 6 weeks compared with 9 months. 

Asthma implies reversible airways obstruction (peak flow varies by >20%, p 271), with wheeze, dyspnoea, or cough. *Prevalence*: >7% (>20% wheeze at some time); prevalence↑ if: ♂, birth weight↓, +ve family history; bottle-fed, atopy, past bronchopulmonary dysplasia, passive smoking. *Differential*: Foreign body, pertussis, croup, epiglottitis, pneumonia, hyperventilation. *Signs of severe asthma*: (Patients may not be distressed). Too breathless to speak or feed; respirations ≥50 breaths/min; pulse ≥140; peak flow ≤50% predicted. *Life-threatening if*: Peak flow <33% of predicted •Cyanosis •Silent chest •Fatigue or exhaustion •Agitation/reduced level of consciousness.

Treatment if >5yrs old: BTS guidelines •Try to avoid provoking factors •Check inhaler technique •Address all fears •Have self-management plan •Check compliance •Give a peak flow meter •Start one step above that indicated by severity, gain rapid control (and the patient's confidence), then step down a rung •Double dose of inhaled steroid if control falters, or there is URTI •Rescue prednisolone may always be needed (≤40mg/day, halved if <4yrs old).

Step 1: Try occasional β-agonists via pMDI. † If needed >daily, add step 2.

Step 2: Add inhaled steroid (eg fluticasone 50–200μg/12h or budesonide 100–400μg/12h) or cromoglicate, eg 20mg/8h as inhaled powder.

Step 3: Add inhaled salmeterol; or† fluticasone (1000μg/12h+large vol. spacer).

Step 4: Add trial of: •Theophylline (eg Slo-Phyllin® <12mg/kg/24h po) or: •Long-acting β-agonist tablet, or •Inhaled ipratropium/oxitropium.

Step 5: Add prednisolone tablets eg 5–10mg po, as a once daily dose.

Dose examples: β-agonists: Sugar-free salbutamol is 2mg/5mL; eg 5mL/8h (0.1 mg/kg/8h if <2yrs). Inhalers are far more preferable: terbutaline 250μg per 4–6h (1 puff)—eg with spacer until 8yrs old. Salmeterol inhaled: eg 50μg per 12h; *Xanthines*: Theophylline, 5mg/kg/8h po (syrup = 60mg/5mL).

Try to monitor levels; bioavailability varies between brands. Capsules are 60, 125, or 250mg. Their enclosed granules may be spread on soft food.

NB: beware growth retardation from steroids (including, rarely, from inhalers: inhaled fluticasone has been found to be least problematic in this regard).

Treatment of severe asthma ►A calm atmosphere is beneficial. *Give these treatments if life-threatening signs are present (above) or the patient is not improving 15–30min after treatment has started.

- | | |
|--------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------|
| 1 Sit the child up; high-flow 100% O ₂ | 6 Oximetry + CXR if S _A O ₂ ≤92% |
| 2 Salbutamol: 5mg O ₂ -nebulized in 4mL 0.9% saline (2.5mg if <2yrs) ± 5μg/kg slow iv | 7 Treat any pneumonia |
| 3 Prednisolone 1–2mg/kg po (max 40mg) ± hydrocortisone 5mg/kg iv over 3min | 8 Chart peak flow before & after each nebulizer treatment; for normal values, see opposite |
| 4* Aminophylline 5mg/kg iv over 20min (omit if already on a xanthine); then iv aminophylline (opposite). Do levels | 9 Repeat nebulizer as needed, eg at 30min, 1h, 2h, 3h, & 4h |
| 5* Hydrocortisone 100mg iv/6h; add ipratropium 0.125–0.25mg to nebulizer | 10 Take to ITU if falling peak flow, exhaustion, confusion, feeble respirations, or coma |

Before discharge ensure: •Good inhaler technique •Has had discharge regimen for >24h •Taking inhaled steroids and oral soluble prednisolone. •Written management plan •Follow-up by GP in 1 week and in clinic in <4 weeks.

Pitfalls ►Faulty inhaler technique. Watch the patient operate her device. •Reluctance to diagnose until a serious attack occurs. Formerly, 50% of diagnoses were made after >15 visits to the GP. Things are now improving. •Inadequate perception of, and planning for, the severe attack. •Unnoticed, marked diurnal variation in airways obstruction. Always ask about nocturnal waking: it is a sign of dangerous asthma. •Being satisfied with less than total symptom control. •Forgetting to start prophylaxis—and not using oral prednisolone early.

Peak flow^{*} (litres/min) in normal boys and girls (5–18 years).

Height (cm)	Mean	Third centile	Height (cm)	Mean	Third centile
100	110		150	360	300
113	160	100	155	400	320
120	210	140	160	420	350
125	240	160	165	450	370
130	260	190	170	470	400
135	290	220	175	500	430
140	310	240	180	520	450
145	350	270	185	550	470
			190	570	500

Which inhaler device is best if <5yrs old? NICE guidelines

- Pressurized metered dose inhalers (pMDI) with spacer system are best for routine use in stable asthma for both steroids and bronchodilators.
- If this is inadequate, nebulizers may be considered if 3–5yrs old. A dry powder inhaler may also be considered.
- Choice of devices should be governed by specific individual need and the likelihood of good compliance.

www.nice.org.uk

Guidelines for continuous IVI of aminophylline—in mg/kg/h after a loading dose[†] of 5mg/kg iv; multiply by 0.85 for the dose of theophylline

- Aim for plasma level of 10–20µg/mL (55–110µmol/L). Serious toxicity (BP↓, arrhythmias, cardiac arrest) can occur at levels ≥25µg/mL.

	Aminophylline (mg/kg/h)
Infants <6 months	0.47
6 months–9 years	1.0
Children 10–16 yrs	0.8

[†]Omit loading dose if previous theophyllines have been taken. If the plasma theophylline concentration is known and is subtherapeutic, an additional loading dose may be given. Increasing the dose by 1mg/kg causes an increase in serum theophylline of ~2µg/mL).

Erythromycin ↑aminophylline's half-life, as do cimetidine, ciprofloxacin, propranolol, and contraceptive steroids. Drugs which decrease the half-life: phenytoin, carbamazepine, barbiturates, and rifampicin. Adjust the dose according to plasma concentrations.

^{*}pMDI=press-and-breathe pressurized meter dose inhaler—as recommended by NICE in 002 'Inhaler devices for ... children...5–15yrs'. ►Monitor inhaler use & evolving needs.

Abdominal pain

Acute abdominal pain Children often have difficulty in localizing pain, and other factors in the history may be more important. Pointers may be:

- Hard faeces suggest that constipation is the cause (p 206).
- In those of African origin, suspect sickle-cell disease (do sickling tests).
- Do tuberculin test (OHCM p 568) if travel or other factors suggest TB risk.
- In children with pica (p 206), do a blood lead level.
- Abdominal migraine is suggested by periodic abdominal pain with vomiting especially if there is a positive family history.
- If any past UTIs, suspect GU disease (eg renal colic or hydronephrosis).

Common physical causes Gastro-enteritis, UTI, viral illnesses (eg tonsillitis associated with mesenteric adenitis), and appendicitis.

Some rarer causes Mumps pancreatitis; diabetes mellitus; volvulus; intussusception; Meckel's diverticulum; peptic ulcer; Crohn's/ulcerative colitis, Hirschsprung's disease; Henoch-Schönlein purpura and hydronephrosis. Consider menstruation or salpingitis in older girls.

► In boys always check for a tortured testis.

Investigations ► Always microscope and culture the urine. Others: consider plain abdominal x-ray; ultrasound; FBC; ESR; IVU, barium studies.

Appendicitis (OHCM p 464) is rare if <5yrs, but perforation rates are high in this group (nearing 90%). Think: how can I tell this from other causes of abdominal pain? • Clues in the history: increasing pain in right lower quadrant, no previous episodes, anorexia, slight vomiting, absence of cough and polyuria. • Examination hint: fever and pulse ↑ are likely; if the child appears well and can sit forward unsupported, and hop, appendicitis is unlikely. • Tests are unlikely to help when the clinical picture is uncertain.

Gastro-oesophageal reflux/oesophagitis Presents with regurgitation, distress after feeds, apnoea, pneumonia, failure to thrive, and anaemia. **Tests:** Endoscopy and evaluation with an oesophageal pH probe is more reliable than barium studies. Ultrasound is not much help. **Treatment:** Reassurance; avoid over feeding (a common cause); lying on the front may help children, but avoid in infants, as this predisposes to cot death (such infants may be helped by lying on the left side). Drugs may be needed, eg an antacid + sodium/magnesium alginate, eg for term infants, Infant Gaviscon® dual dose sachets, 1 dose mixed with 15mL boiled (cooled) water. This paste is given by spoon after each breast feed. If bottle fed, give the dose dissolved in feeds. Children >4.5kg may have a whole dual dose sachet. Carobel® thickens the feeds. If this fails, some experts use domperidone ± omeprazole (or similar). Most resolve by 6–9 months; if not, consider fundoplication, eg if there is failure to thrive, severe oesophagitis, apnoea, or bleeding.

Abdominal distension

Always remember acute GI obstruction as a cause; also consider:

Air:	Ascites:	Solid masses:	Cysts:
Faecal impaction	Nephrosis	Neuroblastoma	Polycystic kidney
Air swallowing	Hypoproteinaemia	Wilms' tumour	Hepatic; dermoid
Malabsorption	Cirrhosis; ccf	Adrenal tumour	Pancreatic

Hepatomegaly Infections: many, eg infectious mononucleosis, CMV.

Malignancy: leukaemia, lymphoma, neuroblastoma (see below).

Metabolic: Gaucher's and Hurler's diseases, cystinosis; galactosaemia.

Others: sickle-cell disease, and other haemolytic anaemias, porphyria.

Splenomegaly All the above causes of hepatomegaly (not neuroblastoma).

Neuroblastoma This is the most common extracranial solid tumour that occurs in early childhood: it is highly malignant and derived from sympathetic neuroblasts. It is thought to be an embryonal neoplasm, presenting with decreasing frequency from birth to 5yrs of age.^{1,2} *Prevalence:* 1:6000–1:10,000. It is the most common solid tumour in the under-5s. *The patient:* is likely to present with abdominal swelling. *Metastatic sites:* Lymph nodes, scalp, bone (causing pancytopenia and osteolytic lesions). In 92%, urinary excretion of catecholamines (vanillylmandelic and homovanillic acids) will be raised. *Treatment:* Refer to special centre. Excision (if possible) and chemotherapy (eg cyclophosphamide and doxorubicin). *Prognosis:* This is adversely influenced by genetics (pseudodiploid karyotypes, chromosome 1p deletions, N-myc gene amplification), less mature catecholamine synthesis and excretion, and age. Those <1yr old do best, and if stage I and II disease, spontaneous remission may occur in 25%. If older, the disease is often disseminated by the time of presentation. *Prevention:* Pre-morbid screening by looking for excretion of catecholamines in the urine detects disease early, but appears not to save lives.^{1,2}

Recurrent abdominal pain At least 10% of children >5yrs suffer recurrent abdominal pains interfering with normal activities. The question is: is there organic disease? No cause is found in most, if looked for, but this should neither encourage complacency (you may delay a diagnosis of *Crohn's* (OHCM p 234) or *peptic ulcer*) or lead to an over-zealous diagnosis of underlying psychological problems (now thought to be less important—but do consider it: who is present when the pain starts; what, or who, makes the pain better?). NB: long-term follow-up indicates a 4-fold ↑risk of psychological problems manifesting in adult life.

Consider these causes: gastro-oesophageal reflux (above), small bowel dysmotility, gastritis, duodenitis, carbohydrate malabsorption (eg lactose, sorbitol), abdominal migraine.

Who to investigate: There are no hard rules. Be suspicious if the pain is unusual in terms of localization, character, frequency, or severity.

Anaemia

The clinical problem You have the results of a full blood count, showing a low Hb ($<11\text{g/dL}$, p 292, the WHO criterion). How should you proceed?

- 1 Take a history (esp travel, diet, ethnic origin); examine the child.
- 2 $\text{mcv} < 70\text{fl}$: suspect iron deficiency or thalassaemia. Features suggesting iron deficiency anaemia (IDA): poor diet, low socio-economic class, bleeding, stomatitis, koilonychia. Features suggesting thalassaemia: of Mediterranean or SE Asia extraction, short stature, muddy complexion, icteric sclerae, distended abdomen (from hepatosplenomegaly), bossed skull, prominent maxillae (from marrow hyperplasia).
- 3 $\text{mcv} > 100\text{fl}$: suspect \downarrow folate (malabsorption, p 185; phenytoin), $\downarrow\text{B}_{12}$ (breast milk from a vegetarian, \downarrow intrinsic factor, malabsorption), or haemolysis.
- 4 $\text{mcv} 81\text{--}97\text{fl}$ (normocytic), suspect haemolysis, or marrow failure (transient, after infections, or chronic, eg thyroid, kidney, or liver failure, or malignancy). Causes of aplasia: chloramphenicol; Diamond–Blackfan syndrome (p 744); Fanconi's anaemia (p 748).
- 5 Is the wcc or differential abnormal? Eosinophilia ($>400 \times 10^9/\text{L}$) + anaemia = hookworms until proved otherwise, in the Tropics.
- 6 Next look at the ESR/CRP. This may indicate some chronic disease.
- 7 Do a film (OHCM, p 626) + reticulocyte count \pm thick films for malaria. Hypochromic microcytic RBCs \approx IDA; target cells \approx liver disease or thalassaemia. Transferrin for IDA; sickling tests + Hb electrophoresis for thalassaemia and sickle-cell anaemia (do in Afro-Caribbeans); B_{12} ; red cell folate.
- 8 Prevention: no cows' milk if $<1\text{yr}$; if formula-fed, use iron-fortified; wean at 4–6 months. Adequate vitamin C intake; iron supplements if premature.

Iron deficiency (20% of the world's population). Despite fortification of breakfast cereals, this is common (12% of white children and 28% of Asian children admitted to hospital). The chief behavioural effect is that iron deficient babies are less happy than others. Dietary causes are the most common—eg poverty, lack of education, or coeliac disease. In recurrent IDA, suspect bleeding (eg Meckel's diverticulum or oesophagitis). Treat with ferrous fumarate syrup ($140\text{mg}/5\text{mL}$), $2.5\text{--}5\text{mL}/12\text{h}$ po (if $<6\text{yrs}$ old; for prems: $0.6\text{mL}/\text{kg}/\text{day}$), or Plesmet®/Sytron®. Warn of the dangers of overdosage. Aim for a rise in Hb of $>1\text{g}/\text{month}$ (do reticulocyte count after 2 months). *Iron deficiency without anaemia*: ►Don't fall into the trap of thinking that if he's not anaemic he's not Fe deficient. CNS iron levels fall before RBC mass. If in doubt, check ferritin. Treating low ferritins may improve: •Memory •Lassitude •Developmental delay •Mood •Cognition—not only in toddlers but in adolescent girls, facing the demands of puberty and menstruation. NB: pica is a sign of iron deficiency.

Haemolysis Get expert help, and try to provide the expert with sufficient information to answer these 4 questions:

- Any evidence of \uparrow RBC production (polychromasia, reticulocytosis)?
- Is there decreased RBC survival? (Bilirubin \uparrow , haptoglobins \downarrow)
- Is there intravascular haemolysis? (Haemoglobinuria)
- Is there an inborn error of metabolism (eg G6PD deficiency), spherocytosis, or is the defect acquired (usually with +ve Coombs' test)?

Sickle-cell disease OHCM p 642. *Pain relief*. Try warmth, hydration, ibuprofen $5\text{mg}/\text{kg}/6\text{h}$ po, or codeine phos $1\text{mg}/\text{kg}/4\text{--}8\text{h}$ po up to $3\text{mg}/\text{kg}/\text{day}$ or Oramorph®, load with $0.4\text{mg}/\text{kg}$ po, then $0.3\text{mg}/\text{kg}/3\text{h}$ + MST® $1\text{mg}/\text{kg}/12\text{h}$ for background analgesia; this may be preferred to iv morphine, eg $0.1\text{mg}/\text{kg}$.

Patient-controlled analgesia: Put morphine $1\text{mg}/\text{kg}$ in 50mL 5% dextrose; try $1\text{mL}/\text{h}$ with self-delivered extra boluses of 1mL as needed. Do respiration & sedation score every $\frac{1}{4}\text{h}$ + $\text{S}_\text{A}\text{O}_2$ if chest or abdominal pain.

Purpura in children

If a child is ill with purpura he or she should be presumed to have meningococcaemia (and be given penicillin, p 258), leukaemia or disseminated intravascular coagulation (investigate initially with a visual non-automated blood film and wcc). If the child is well, and there is no history of trauma, the cause is likely to be Henoch-Schönlein purpura or idiopathic thrombocytopenic purpura, readily told apart by the normal platelet count in the former. (Aplastic anaemia is very rare.)

Idiopathic thrombocytopenic purpura *Presentation:* Acute bruising, purpura, and petechiae. Less commonly there is bleeding from gums, nose or rectum. If this is present to any large extent, or there is lymphadenopathy, hepatosplenomegaly, or pancytopenia, another diagnosis is likely. *Tests:* The peripheral blood film usually shows only simple thrombocytopenia. ITP may be a response to a virus, and it is worth looking for congenital CMV if the patient is <1yr old. Also look for Epstein-Barr virus and erythrovirus (formerly parvovirus B19). There may also be a lymphocytosis. It is not necessary to do a marrow biopsy, unless:

- Unusual features are present, eg equivocal or abnormal cells on a film.
- Platelet count is $<30 \times 10^9/L$ or the count is not rising after ~2 weeks.
- Treatment is contemplated with steroids or immunoglobulins.

Platelet antibody tests are unhelpful. If SLE possible, do DNA binding. CT to exclude cerebral bleed, eg if there is headache or CNS signs.

Natural history: The course may be gradual resolution over ~3 months, or the course may be more chronic (>6 months in 10–20%). The chronic form is compatible with normal longevity, and normal activities of daily living, provided contact sports are avoided.

Management: Consider admitting to hospital if: • Unusual features, including excessive bleeding. • Social circumstances. • There are rowdy siblings at home who might engage in physical badinage (the risk of intracranial haemorrhage is <1%).

Splenectomy may benefit $\frac{2}{3}$ of those with chronic ITP if >6yrs old, and have had ITP for >1yr. NB: Platelet transfusion is only indicated if very severe haemorrhage (and during splenectomy). Steroids and immunoglobulin are now not used (unfavourable risk/benefit ratio). Follow-up of 92 children with chronic ITP revealed only one death from bleeding, and this was associated with a major physical assault.

Henoch-Schönlein (anaphylactoid) purpura (HSP) presents with purpura (purple spots/nodules which don't disappear on pressure), intradermal bleeding eg over buttocks and extensor surfaces (\pm urticaria). $\sigma/\phi >1:1$. *Other features:* • Nephritis (with crescents, in $\frac{1}{3}$ of patients—an IgA nephropathy—we could think of HSP as being a systemic version of Berger's syndrome, *OHCM* p 720) • Joint involvement common • Abdominal pain (as if 'acute abdomen') • Intussusception.

The fault lies in the vasculature (platelets are normal). It may follow respiratory infection, and is typically selflimiting after a few months. Complications (worse in adults): massive GI bleeding, ileus, haemoptysis (rare), and renal failure (rare). One option in HSP nephritis (not usually required) is high-dose steroids + cyclophosphamide; this reduces proteinuria (a risk factor for renal insufficiency in HSP).

medline:11359020

Upper respiratory infection URTI OHCM p 547, sore throats. p 556

Stridor and epiglottitis ►► Acute stridor may be a terrifying experience for children and this fear may lead to hyperventilation, which makes symptoms worse. Causes: p 558. The leading causes to be distinguished are viral croup and epiglottitis (rare in the UK since vaccination): see table opposite.

Investigations: A lateral neck x-ray may show an enlarged epiglottitis (but do not insist on x-rays at the expense of upsetting the child).

Croup (Acute laryngotracheobronchitis) *Epidemics:* Autumn & spring. *Causes:* Parainfluenza virus (types 1, 2, 3), respiratory syncytial virus, measles (rare). Symptoms are caused by subglottic oedema, inflammation, and exudate.

Management of croup: Mostly at home; ask parents to watch for respiratory rate, fatigue, and restlessness. Anecdotal evidence says that a warm, humid environment is helpful, but mist tents have lost favour as they frighten children, and subsequent hyperventilation increases distress. In hospital, aim for minimal interference and careful observation (TPR; $S_{A}O_2$) by experienced nurses. Watch for restlessness, rising pulse and respiration rate, chest-wall indrawing, fatigue/drowsiness. This will prompt intubation under general anaesthesia. If severe, admit to hospital, and buy time with nebulized epinephrine (=adrenaline) 1:1000 (one dose of 1mL). Nebulized budesonide also helps—loading dose: 2mg, then 1mg/12h eg as Pulmicort®. Oral dexamethasone, 0.15mg/kg (by mouth or injected) is equally effective, and cheaper.

Management of suspected epiglottitis ►► Avoid examining the throat. This may precipitate obstruction. Once in hospital, summon the most experienced anaesthetist. Ask her to make the diagnosis by laryngoscopy. If the appearances are those of epiglottitis (a cherry-red, swollen epiglottis), electively intubate, before obstruction occurs. (A smaller diameter endotracheal tube than normal for that age may be needed—so don't pre-cut all your tubes!) Then do blood cultures. The cause is usually *Haemophilus influenzae* type b and strains resistant to ampicillin (and less so chloramphenicol) are prevalent, so the only safe initial treatment is a 3rd generation cephalosporin (eg cefotaxime 1–2g/8h iv adults, 25–50mg/kg/8h iv children). Hydrocortisone (1–2mg/kg/6h iv) is often given, but is not of proven value. Expect to extubate after about 24h (longer if the diagnosis turns out to be a staph laryngotracheobronchitis, when flucloxacillin 50mg/kg/6h iv should also be given).

Diphtheria is caused by the toxin of *Corynebacterium diphtheriae*. It usually starts with tonsillitis ± a false membrane over the fauces. The toxin may cause polyneuritis, often starting with cranial nerves. Shock may occur from myocarditis, toxæmia, or involvement of the heart's conducting system. Other signs: dysphagia; muffled voice; bronchopneumonia; airway obstruction preceded by a brassy cough (laryngotracheal diphtheria); nasal discharge with an excoriated upper lip (nasal diphtheria). If there is tachycardia out of proportion to fever, suspect a toxin-induced myocarditis. Monitor with frequent ECGs. Motor palatal paralysis also occurs.

Diagnosis is by swab culture of material below the pseudomembrane.

Treatment: Diphtheria antitoxin: 10,000–30,000u IM (any age) and erythromycin. Give contacts 7 days' erythromycin syrup: <2yrs old 125mg/6h po (500mg po 6h if >8yrs) before swab results are known. UK vaccination has been effective.

Prevention: Isolate until 3 –ve cultures separated by 48h. Routine vaccination: p 209. There is a resurgence of diphtheria in Moscow and St Petersburg.*

*Those born before 1942 visiting Russia or the Ukraine who have close local contacts need a primary low dose course. Give a booster (0.5mL of Diftavax®; contains tetanus toxoid too) to diphtheria contacts whose primary immunization was >10yrs ago. Explain that it may not work.

Viral croup vs

Onset over a few days
Stridor only when upset
Stridor sounds harsh
Voice hoarse
Barking cough
Likely to be afebrile
Can swallow oral secretions

Epiglottitis (supraglottitis)

Sudden onset
Continuous stridor
Stridor softer, snoring
Voice muffled/whispering
Cough not prominent
Toxic and feverish (eg $T^{\circ} > 39^{\circ}\text{C}$)
Drooling of secretions

Note: the distinction may not be clear cut. If in doubt, admit to hospital.

Lower respiratory infections

Each year ~3 million infants die from acute respiratory infections (WHO).

► If severely ill, think of staphs, streps, TB, and HIV (pneumocystosis, *OHCM* p 160). In chronic cough think of: • Pertussis • TB • Foreign body • Asthma.

Acute bronchiolitis is the leading lower respiratory tract infection in infants. Coryza precedes cough, low fever, tachypnoea, wheeze, apnoea, intercostal recession ± cyanosis. *Typical cause:* Winter epidemics of respiratory syncytial virus. *Diagnosis:* immunofluorescence of nasopharyngeal aspirates. *Other causes:* mycoplasma, parainfluenza, adenoviruses. Those <6 months are most at risk. *Signs prompting admission:* feeding difficulties, tachypnoea >50 breaths/min, apnoea, dehydration, exhaustion. *Tests:* CXR (hyperinflation); blood gases/oximetry; FBC. *Treatment:* If severe, give O₂; nasogastric feeds. 5% need ventilating (mortality ≈1%; 33% if symptomatic congenital heart disease). Antibiotics & steroids are generally not indicated. The rôle of ribavirin is uncertain. medline:11316109 &: 9099772

Pneumonia Signs: fever, malaise, feeding difficulties, tachypnoea, and cyanosis. Older children may have typical lobar pneumonia, with toxæmia and pleural pain. Auscultation: creps; bronchial breathing (easy to miss). CXR: consolidation. Cavitation suggests TB or staphs. Take samples (blood; MSU) before starting 'blind' antibiotics, eg penicillin G 25mg/kg/4h IV + erythromycin eg as sugar-free solution (250mg/5mL)—5mL/12h PO for babies ≤2yrs, 10mL/12h from 2–8yrs; if severely affected, give this dose 6-hourly. Alternative: amoxicillin. High-flow O₂ may be needed. If compliance is a problem, once daily azithromycin may be the answer: 10mg/kg, ie 200mg (5mL) if 15–25kg, PO for 3d. *Causes:* *Pneumococcus*, *Haemophilus*, *Staphylococcus*, *Mycoplasma* (hence choice of erythromycin), TB, viruses.

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TB Suspect if: overseas contacts, HIV+ve; odd CXR. *Signs:* Anorexia, low fever, failure to thrive, malaise. Cough is common, but may be absent. *Diagnosis:* Tuberculin tests (*OHCM* p 568); culture and Ziehl–Neelsen stain of sputa (×3) and gastric aspirate. CXR: consolidation, cavities. Miliary spread (fine white dots on CXR) is rare but grave. *Treatment:* Seek expert help. 6-month supervised regimen: isoniazid 15mg/kg PO 3 times a week + rifampicin 15mg/kg/PO ac 3 times a week + pyrazinamide (first 2 months only) 50mg/kg PO 3 times a week. Monitor U&E and LFT before and during treatment. Stop rifampicin if the bilirubin rises (hepatitis). Isoniazid may cause neuropathy (give concurrent pyridoxine). ► Explain the need for prolonged treatment. Multiple drug resistance: *OHCM* p 567.

Whooping cough (*Bordetella*) *Signs:* Bouts of coughing ending with vomiting (± cyanosis), which are worse at night, and after feeding. The whoop (not always present) is caused by inspiration against a closed glottis. Differentiation from pneumonia, asthma, bronchitis, and bronchiolitis is aided by the (usual) absence of a fever >38.4 °C and wheeze. *Peak age:* 3yrs. In the UK, the illness is usually mild, with ~1% needing admission (eg with secondary pneumonia). *Diagnosis:* Culture is unsatisfactory: organisms often die on the way to the lab. Fluorescent antibody testing of nasopharyngeal aspirates is specific but insensitive. Absolute lymphocytosis is common (sometimes very high). *Incubation:* 10–14 days. *Complications:* Prolonged illness (>3 months). Coughing bouts may cause petechiae (eg on cheek), conjunctival, retinal & CNS haemorrhage, apnoea, inguinal hernias, and lingual frenulum tears. Deaths may occur (esp. in infants), as may late bronchiectasis. *Treatment/prevention:* Salbutamol and steroids don't help much. Erythromycin is often used in those likely to expose infants to the disease (benefit unproven). Admit if <6 months old (risk of apnoea). *Vaccine:* p 209.

The 'chesty' infant

Many children with cough and wheeze do not fit into the categories opposite, and are too young for a diagnosis of asthma to be made with confidence. These infants often end up being treated with escalating bronchodilator therapy with frequent courses of antibiotics against uncultured organisms. While it is true that asthma can begin in infancy, most of these chesty infants do not have asthma—but we tend to prescribe 'just in case'. As the natural history of symptoms is to vary from hour to hour, sometimes we *appear* to be successful. NB: *viral wheeze* may be the appropriate label here: it is a non-atopic disorder. An alternative diagnosis may be *altered awareness of minor symptoms*. medline:11410189

The rôle of passive smoking is under investigation. 'Happy wheezers' (ie undistressed) probably need no treatment, but if chest symptoms start very early in life, a sweat test is needed to rule out cystic fibrosis. Between these two ends of the spectrum of 'chestiness' lie those who clearly need some help. These may benefit from relatively high-dose inhaled steroids, given regularly for 8 weeks or so. medline:9596586

Assess benefit by reference to reduction in sleep disturbance.

Aim to engage in a constructive dialogue with parents so that they understand that treatment is often unsuccessful, but that their child is unlikely to come to harm, while he or she is 'growing out of it'.

If cough is a chronic problem, exclude serious causes (eg TB; foreign body; asthma) and reassure. There is no good evidence that brand name cough medicines are better than placebo—so avoid them.

Renal failure and some renal diseases

Acute renal failure Essence: an acute severe fall in glomerular filtration rate (GFR) leading to a rapidly rising K^+ and urea often with anuria ($<100\text{mL/day}$) or oliguria ($<200\text{mL/m}^2/\text{day}$) and $\uparrow\text{BP}$. *Causes:* Acute tubular necrosis (ATN); GU obstruction; toxins (eg sulfonamides); glomerulonephritis (GN, *OHCM* p 256); haemoglobinuria; myoglobinuria; (*OHCM* p 271). *Causes of ATN:* •Crush injury •Burns •Dehydration •Shock •Septicaemia •Malaria.

Plasma biochemistry: $K^+\uparrow$, urea \uparrow , $\text{PO}_4^{3-}\uparrow$; $\text{Ca}^{2+}\downarrow$, $\text{Na}^+\downarrow$, $\text{Cl}^-\downarrow$.

MSU: Are there red cell casts (=GN)? If no RBCs seen but Labstix +ve for RBCs, consider haemo/myoglobinuria (*OHCM* p 271).

Other tests: ECG, serum and urine osmolality, creatinine, acid–base state, PCV, platelets, clotting studies (DIC), C3, ASO titre, ANA (antinuclear antibody).

Radiology: ►Arrange prompt abdominal ultrasound. Are the ureters dilated (eg stones)? If so, urgent surgery may be required.

Treatment: Remove or reduce the cause, promptly.

- Treat shock and dehydration (p 250)—then:
- If the urine/plasma (u/p) osmolality ratio is >5 the kidneys concentrate well; the oliguria should respond to rehydration. If the u/p ratio is low, try for a diuresis: furosemide 1.5mg/kg iv slowly, maximum 20mg/day .
- Monitor BP. If $\text{BP}\uparrow\uparrow$: nitroprusside (p 281).
- 24h fluid requirement: Avoid overhydration. Replace losses + insensible loss ($12\text{--}15\text{mL/kg}$). Aim for weight loss ($0.5\%/ \text{day}$).
- Give no K^+ . Monitor ECG. Tall T-waves and QRS slurring prompt urgent lowering of K^+ , with iv salbutamol $4\mu\text{g/kg}$ or 10mg nebulized (2.5mg if $<25\text{kg}$). A less easy to use alternative is glucose (4g/kg) with soluble insulin ($1\text{u}/4\text{g}$ of glucose) ivi over 2h. medline:8129434. Also consider Resonium A® 0.5g/kg PO and calcium gluconate (10% , 0.5mL/kg iv over 10min; monitor ECG: stop ivi if heart rate \downarrow) to counteract electrophysiological effect of hyperkalaemia.
- High energy, protein limited diet to slow catabolism.
- Dialysis indications (a guide, only): $K^+ >7\text{mmol/L}$; urea $>40\text{mmol/L}$; $\text{HCO}_3^- <13\text{mmol/L}$; severe hypertension; overhydration.
- Improvement is ushered in with a diuretic phase.

Haemolytic uraemic syndrome Essence: Microangiopathic haemolytic anaemia, thrombocytopenia, renal failure, and endothelial damage to glomerular capillaries. *Typical age:* 3 months to 3yrs. Epidemics may occur from *E coli* Verocytotoxin. Diarrhoea is usual but not always present.

Other causes: Shigella, HIV, SLE, drugs, tumour, scleroderma, $\text{BP}\uparrow$.

Clinical features Colitis \rightarrow haemoglobinuria \rightarrow oliguria \pm CNS signs \rightarrow encephalopathy \rightarrow coma. LDH \uparrow , WCC \uparrow , Coombs' -ve. PCV \downarrow . Fragmented RBCs.

Mortality. 5–30%. A few have many relapses—to form a condition rather like thrombotic thrombocytopenic purpura (TTP).

Treatment: Seek expert advice. Treat renal failure (above). There is no evidence that fibrinolytic agents or anticoagulation help. Relapses in TTP may be preventable by steroids, splenectomy, or vincristine.

Chronic renal failure *Causes:* Congenital dysplastic kidneys, pyelonephritis, glomerulonephritis; chronic infection, reflux nephropathy—also any cause of acute renal failure which leads to cortical necrosis. ►Monitor growth, BP, U&E, Ca^{2+} , PO_4^{3-} .

The Child: Weakness, tiredness, vomiting, headache, restlessness, twitches, $\text{BP}\uparrow$, hypertensive retinopathy, anaemia, failure to thrive, seizures, and coma.

Treatment: (See box). Talk with experts about haemodialysis & transplants.

Chronic renal failure: metabolic and other issues

Get a dietitian's help. Calorie requirements may not be met if nausea and vomiting are problems. Eggs and milk may be appropriate (high biological protein value). Provide protein at a level of 2.5g/kg/24h. Vitamin drops may also be needed. Nasogastric or gastrostomy tube feedings may have a rôle. Growth hormone therapy combined with optimal dialysis improves linear growth.

Acidosis is common needing no treatment if serum bicarb is ≥ 20 mmol/L. Sodium bicarbonate tablets (600mg=7.14mmol) or 1.26% solution (0.15mmol of sodium and bicarb in 1mL) may be used, 1–2mmol/kg/day po for babies <2yrs old; if older, 70mmol/m²/day). SE: Na⁺ causes BP↑.

Renal osteodystrophy: A bone disease resulting from poor mineralization due to renal failure, causing growth retardation, muscle weakness, bone pain, skeletal deformities, and slipped epiphyses. It is like rickets and osteomalacia (↓parathormone [PTH] release causes ↓bone turnover). If glomerular filtration rate falls to $\leq 25\%$ of normal, compensatory mechanisms to enhance phosphate excretion fail; the resulting hyperphosphataemia promotes hypocalcaemia, so PTH rises, which enhances bone resorption to release calcium in an attempt to correct hypocalcaemia. PTH↑ leads to marrow fibrosis (osteitis fibrosis cystica). Also, the failing kidneys cannot convert adequate amounts of 25-hydroxycholecalciferol to active 1,25-dihydroxycholecalciferol, so GI calcium absorption falls, so worsening hypocalcaemia. Management: Aim to normalize calcium and reduce the associated hyperphosphataemia, eg with PTH of 200–400pg/mL.

Hyperphosphataemia may be controlled by enhancing faecal phosphate excretion with calcium carbonate taken immediately before food, which binds phosphate. Typical dose: <1yr: 120mg before food (3 times a day) 1–5yrs: 300mg before meals; 5–12yrs 600mg before meals; 12–18: 1.25g before meals. A dose may also be taken before bed.

If Ca²⁺ is low despite correcting serum phosphorus, give 1,25-dihydroxycholecalciferol (calcitriol), eg 15nanograms/kg/24h po. The dose may be increased in increments of 5ng/kg until Ca²⁺ and alk phos are normal, PTH level is reduced to 200–400pg/mL. X-ray evidence of healing rickets may also be sought. Then reduce the dose. SE: ↓renal function, hypercalcaemia, and hyperphosphataemia. Because normal bone requires adequate levels of PTH to promote bone modelling, oversuppression of PTH must be avoided to avoid adynamic osteodystrophy.

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Anaemia is common, and is primarily the result of inadequate renal production of erythropoietin (± poor iron and folic acid intake). A typical Hb is 6–9 g/dL. Do not transfuse, as this suppresses erythropoietin production. Subcutaneous erythropoietin may be indicated in pre-dialysis and peritoneal dialysis patients, and intravenously to patients on haemodialysis.

Hypertensive emergencies: Get expert help. Use sodium nitropruside 0.5–1µg/kg iv then 1–8µg/kg/min ivi by pump to allow precise control of ivi rate. Protect from light. Monitor BP continuously, ↑dose slowly to the required level. CI: severe hepatic impairment. Withdraw over >10–30min to prevent rebound hypertension. In prolonged use (>1 day), monitor blood and plasma cyanide levels (keep to <38µmol/L and <3µmol/L, respectively). Labetalol is an easier to use alternative, starting dose, eg 0.1–1mg/kg/dose iv, then up to 2mg/kg/h iv. medline:8827760 &: 1417052

Acute nephritis and nephrosis

Acute nephritis Essence: haematuria and oliguria (\pm hypertension and uraemia) produced by an immune mechanism in the kidney. Age peak: 7yrs.

Causes: Often β -haemolytic streptococcus, which may cause a sore throat 2 weeks before an acute glomerulonephritis. Others:

- Henoch–Schönlein purpura
- Toxins, heavy metals
- Berger's disease (OHCM p 720)
- Malignancies
- Infections: viruses; SBE; syphilis
- Renal vein thrombosis

Uncomplicated presentation: Haematuria; oliguria; BP \uparrow (50%); periorbital oedema; fever; GI disturbance; loin pain. **Other presentations:** 1 Hypertensive encephalopathy (restless, drowsy, bad headaches, fits, vision \downarrow , vomiting, coma). 2 Cardiac involvement: gallop rhythm, cardiac failure and enlargement, pulmonary oedema. 3 Uraemia: acidosis, twitching, stupor, coma.

MSU: count RBCs, WBCs, hyaline, granular casts; red cell casts = glomerular bleeding). Phase-contrast detects odd-shaped red cells, signifying glomerular bleeding. This change may not be present at first. 24h urine for protein and creatinine clearance. Check urine culture, and specific gravity (normal range: infants 1.002–1.006; child/adult 1.001–1.035).

Blood tests: Urea \uparrow in $\frac{2}{3}$; ESr \uparrow ; acidosis. Complement (C3) often \downarrow 2–8 weeks after onset (not in Henoch–Schönlein purpura). Find the cause: do ASO titre, antinuclear factor, syphilis serology, blood cultures, virology.

Other tests: Renal ultrasound; renal biopsy (check platelets & clotting pre-op).

Treatment: Restrict protein when in oliguric phase. Give penicillin 10mg/kg/4h iv for the first few days, then po for 3 months to prevent further streptococcal infection. Measure BP often. Treat severe hypertension. If encephalopathy, give nitroprusside 0.5–8 μ g/kg/min ivi (p 281).

Nephrotic syndrome (nephrosis) Essence: oedema, proteinuria (eg 4g/24h), hypoproteinaemia \pm hypercholesterolaemia. In 90% the cause is unknown, but any of the causes of nephritis (above) may also cause nephrosis. Histology: usually minimal change. Symptoms: anorexia, GI disturbance, infections, irritability; then oedema (periorbital, genital), ascites, oliguria.

Urine: Frothy; albuminous \pm casts; Na $^{+}$ \downarrow (secondary hyperaldosteronism).

Blood: Albumin \downarrow (so total Ca $^{2+}$ \downarrow); urea and creatinine usually normal.

Renal biopsy: Reserve this for older children with any of: haematuria, BP \uparrow , urea \uparrow , if protein loss is unselective (ie large molecular weights as well as small), and treatment 'failures'. SE: haematuria; haematoma. medline:9553292

Complications: Pneumococcal peritonitis or other spontaneous infections. Consider pneumococcal vaccination if >2 yrs.

Treatment: Limit oedema with protein-controlled (3g/kg/24h), low Na $^{+}$ diet (<50 mmol/24h). Consider furosemide 1–2mg/kg/24h slow iv/po + spironolactone (1.2mg/kg/12h po). For 'minimal change' give prednisolone, max 1mg/kg/24h po (not if incubating chickenpox!) for 9 weeks, or 1 week after remission-induction. Half the induction dose is then given, eg for 1 month, then tapering. Total length of steroid course: 3–7 months. 2001 medline:11405997

90% respond in 8 weeks. $\sim 30\%$ of responders have no relapse. 10–20% of these are cured after ≤ 4 courses of steroids. The rest may become dependent on steroids: if >0.5 mg/kg/day of prednisolone is needed, growth may be slowed (esp. at adolescence, and agents such as cyclophosphamide may be indicated (2–2.5mg/kg/day po, SE: haemorrhagic cystitis, wcc \downarrow , hair loss, infertility). Alternatives: chlorambucil 0.2mg/kg/day; ciclosporin (nephrotoxic, OHCM p 676). medline:9707852

► Monitor BP. Meticulous control minimizes progression to renal failure.

Steroids in nephrotic syndrome

In nephrotic syndrome, protein leaks from blood to urine through glomeruli, causing hypoproteinaemia and oedema. Before steroids (and antibiotics), many died from infections. Most children with nephrotic syndrome respond to corticosteroids, but many experience a relapsing course with recurrent oedema and proteinuria. Corticosteroids reduce mortality to ~3%, with infection remaining the most important cause of death. NB: steroids cause obesity, poor growth, BP↑, diabetes, osteoporosis, avascular necrosis (hip), and adrenal suppression.

medline:11405997

Steroid-resistant proteinuria and ACE-i

Enalapril has been used in courses of >2yrs (0.2–0.6mg/kg/24h po). In one study, urine protein electrophoresis showed a reduction of 80% and 70% in the total protein and albumin, respectively, after enalapril. Some patients become free of proteinuria. ACE-i are discontinued if renal failure occurs, eg during infections.

medline:11045392 & 10879662

Non-accidental injury (NAI)

This page should be read in conjunction with your local child protection guidelines and the relevant legislation in the country in which you are practicing, eg in England, the Children's Act (one provision of which is that the child's welfare is paramount). Abuse may be physical, sexual, emotional, or by neglect. In *Munchausen's by proxy*, a parent fabricates alarming symptoms in their child to gain attention—and possibly dangerous medical interventions. *Risk factors*. Birth weight <2500g; mother <30yrs; unwanted pregnancy; stress; social class IV and V *Suspect abuse if*: disclosure by child, or:

- Odd story, lacking congruence with injuries
- Odd mode of injury*
- Late presentation, to an unknown doctor
- Odd constellation of signs
- The accompanying adult may not be a parent
- Parental financial problems
- Efforts to avoid full examination, eg after an immersion burn.
- Psychological sequelae (stress; depression) from sexual or emotion abuse.
- Unexplained fractures; injury to buttocks, perineum or face; intracranial haemorrhage; torn lingual frenulum; vitreous/retinal bleeds, hyphaema (p 514), lens dislocation, bulging fontanelle, head circumference \uparrow \pm xanthochromia; \blacktriangleright if in doubt, do cr.* *Other signs*: Cigarette burns; linear whip marks (look for outline of a belt buckle, or the loop of a double electric flex); bruised *non-mobile* baby; signs of suffocation or finger-mark bruising.

Difficult questions Although non-medical, these need addressing because we are not simply technicians, and they influence what we do, how we react to child abuse, and how the rest of society perceives our rôle.

- Could *proving* of abuse be more destructive than the abuse itself?*** Note that even if the answer is yes, society places a duty on us to report abuse.
- Is it better for him to be loved and battered than neither?
- Is help from the extended family more desirable than the law? Is it possible that the parents can grow through crisis, as abuse is discussed, and help given?

Remember that the first aim is to prevent organ damage, murder, and other significant harm. If this is a real danger \blacktriangleright *contact the duty social worker today*—eg for an emergency protection order. Offer help to the parents. Learn to listen, leaving blame and punishment to judges. Find out about local policies and referral routes. Remember that very often your duty is not to diagnose child abuse, but to recognize *possible* abuse, and then to get help.

Sexual abuse This may be prevented by teaching about 'personal safety' and how to say 'No'. Know your local guidelines. Follow them. Inform Social Services. If you do not, ask yourself with whom you are colluding. Forensic specimens (eg pubic hair, vaginal swabs) are to be taken by an expert who knows to be gentle, and to avoid a 'second rape'. Prepubertal venereal disease means abuse until proven otherwise. Does abuse cause psychological harm? Yes, sometimes, but the position is complex. See p 323.

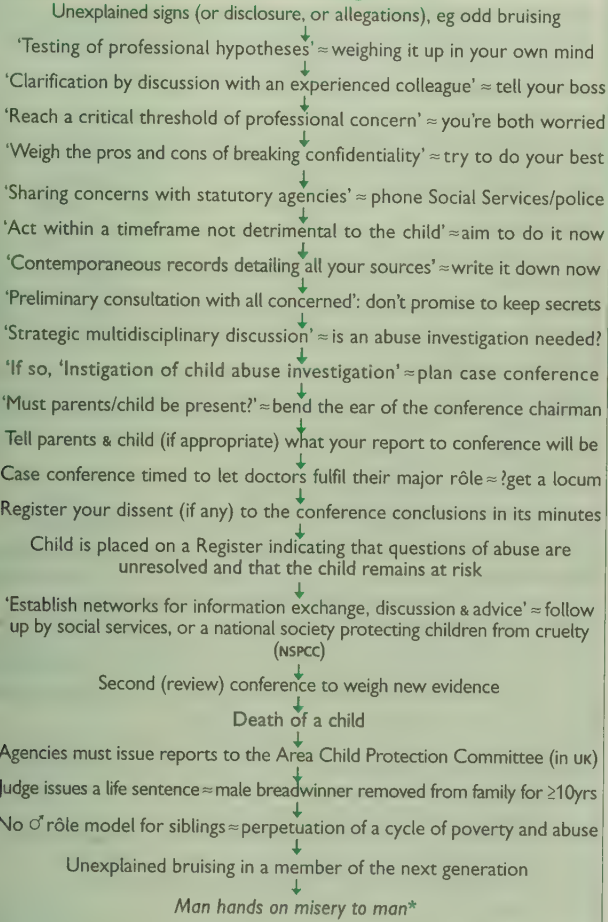
Repertoire of actions in primary care \blacktriangleright After informing Social Services, liaise with health visitor (may be a very helpful source of information) or NSPCC (National Society for the Prevention of Cruelty to Children).

- Admission to a place of safety (eg hospital or foster home).
- Continuing support for parents and protection for siblings.
- Prevention: encourage impulses to be shared, and not acted on.
- Attend a 'case conference' (social worker, health visitor, paediatrician and police) to decide what action to take. NB: it isn't known if conferences are the best way of making such decisions: not all members will know the family.

Other forms of abuse Emotional abuse; neglect; exposure to the effects of domestic violence, alcohol or drug abuse, serious mental illness in carers.

*Look for injuries inconsistent with child's level of development. UK phone no for adolescents in distress: ChildLine 0800 884 444 validated in Arch Dis Child 2000 82 283 ***K Hulme 1984 Bone People, Spiral

A possible sequence of events might be:



Man hands on misery to man.
 It deepens like a coastal shelf.
 Get out as early as you can,
 And don't have any kids yourself.

* Philip Larkin, *This Be The Verse*

Not all our efforts to protect children end thus. Successes are frequent. This sequence is an oversimplification. The sign ≈ is not meant flippantly: it is intended as a short-hand, denoting the exercise of reflection, good judgment, action, and the following of agreed procedures.

Sudden infant death syndrome (sids)

Definition^{6*} 'Sudden death of an infant under 1 year of age, which is unexpected after a thorough case investigation, including a complete autopsy, examination of the death scene, and review of the clinical history.'

► sids is the leading 'cause' of death in infants aged over 1 week old.

Epidemiology *Peak incidence:* 1–4 months; *Risk:* Lower socio-economic classes, passive smokers, males, premature babies, winter, previous sibling affected by sids; co-existing minor upper respiratory infection is common.

There are many theories as to the cause:

medline:1799550 & 129359

Obstructive apnoea:

Central apnoea:

Others: Long Q-T interval*

Inhalation of milk

Faulty CO₂ drive

Overheating; ↑Vagal tone;

Airways oedema ± collapse

Prematurity

Immature diaphragm; Mg²⁺ ↓

Passive smoking

Brainstem gliosis

Viruses; Genes medline:10783475

Sleeping supine ('back to sleep'), preventing overheating, and cigarette smoke exposure are the main areas for intervention/prevention: risk from passive smoking is dose-dependent, and often at least doubles risk. medline:10742373

The face is an important platform for heat loss—and it is known that the incidence of sids is ~5–10-fold higher among infants usually sleeping prone (17-fold higher if sleeping in a room separated from parents): ►so recommend sleeping supine (unless there is gastro-oesophageal reflux, Pierre Robin syndrome, or scoliosis). Advise as follows:

- Do not overheat the baby's bedroom. Aim for a temperature of 16–20°C.
- Do not use too much bedding, and avoid duvets if less than 1 year of age.
- If ill or feverish, consult a GP—do not increase the amount of bedding.
- Have feet come down to the cot's end to avoid migrations under blankets.
- While sleeping, avoid heaters, hot water bottles, electric blankets and hats unless ambient T° is very low. Do wrap up if going out in winter.
- Babies >1month do not need to be kept as warm as in hospital nurseries.

Autopsy is unrevealing; minor changes are common; petechial haemorrhages over pleura, pericardium, or thymus, and vomit in the trachea may be agonal events. *Causes to exclude:* meningococcaemia; epiglottitis; heart defects; murder.

How the GP can help the family on the first day

- A prompt visit to express sympathy emphasizing that no one is to blame.
- Explain about the legal requirement for an autopsy and coroner's inquest. The parents may be called upon to identify the body.
- Bedding may be needed to help find the cause of death.
- Don't automatically suppress lactation (bromocriptine 2.5mg/12h po for week), as continued lactation may be an important way of grieving.
- Many parents will not want anxiolytics, but may want hypnotics.
- Admit a twin sibling to hospital (their risk is increased).

Subsequent help^{**} Advise the parents of likely grief reactions (guilt, anger, loss of appetite, hearing the baby cry). Make sure that the coroner informs you of the autopsy result; take some trouble to explain these to the parents. Offer them a chance to talk to a consultant paediatrician. This can provide helpful reinforcement and encouragement to the parents and yourself. The parents may find an electronic apnoea alarm reassuring in caring for later infants. Ask if they would like to join a self-help group. Programmes exist to prevent a future sid—eg the con1 programme (care of next infant)

*Risk ↑ if Q-T corrected for rate (QT_c) ≥440msec; NB: QT_c = (Q-T)/(R-R) [11056286] [10753246]

**UK Cot Death Helpline (24h) 0845 601 0234; 14 Halkin St, London SW1

Denver developmental screening test

This is a detailed test to consider if results of simple tests are equivocal (p 208). Begin with observation, as children will often spontaneously demonstrate many of the milestones. Then do the specific tests, and then question the parents about what words the child can say. Younger children may be examined while sitting on their mother's lap. Explain to her that the child is not expected to pass every test. Begin with a few very easy tests. Go slowly.

Test materials Ball of red wool, box of raisins, rattle with handle, small bottle, bell, tennis ball, 8 blocks (one inch cubes).

Administering the test Draw a vertical line at the child's chronological age on the charts on p 290–1. If premature, subtract the months premature from the chronological age. Note that:

- Items to be administered are those through which the line passes. A parent may administer a test if the child wishes.
- Failure to pass an item passed by $\geq 90\%$ of children may or may not be abnormal. Note how the child behaves during the test.

Test footnotes (See top corner of test boxes on p 290–1.)

1 When prone lifts head up, using forearm support (\pm hands).

2 No head lag as you pull by the hand from supine to sitting.

3 Child may use a wall or rail for help (but not a person).

4 Throws ball overhand 3 feet to within your reach.

5 Jumps over a distance (eg over an A4 piece of paper).

6 Walk forward, heel within one inch of toe.

7 Bounce a ball. He must catch it. Allow up to 3 trials.

8 Walk backwards, toe within 1 inch of heel.

9 Wave wool slowly before eyes: do eyes move 90° to midline?

10 Grasps rattle when it touches his finger tips.

11 Looks for ball of wool dropped out of sight over table's edge.

12 Child grasps raisin between thumb and index finger.

13 Overhand grasp of raisin between thumb and index finger.

14 'Copy this' (circle). Do not name or demonstrate.

15 'Which line is longer?'; turn upside down and repeat.

16 'Copy this' (cross). Pass crossing lines at any angle.

17 'Copy this' (square). You may demonstrate if he fails.

18 Two arms or two eyes or two legs only count as one part.

19 Name the pictures at the bottom of the chart.

20 'Give the block to Mum'. 'Put it on the table'. No gestures.

21 Answer $\frac{2}{3}$ of: 'What do you do if you are cold/hungry/tired?'

22 Put this on/under/in front of/behind the chair. No gestures.

23 Answer $\frac{2}{3}$: 'Fire is hot, ice is –'. 'Mum is a woman, Dad is a –'. 'A horse is big, a mouse is –'.

24 Define $\frac{5}{8}$ of ball, lake, desk, house, banana, curtain, hedge, pavement. A verbal indication of understanding is passed.

25 Answer $\frac{3}{3}$ of: 'What is a spoon/shoe/door made of?' (No others).

26 Smile, talk or wave, to elicit smile (3 tries). Do not touch.

27 While he plays with a toy, pull it away. Pass if he resists.

28 Child need not be able to tie shoes or button at the back.

► This test is poor at picking up articulatory or mild linguistic deficits—if this is suspected, liaise with speech experts. **NB:** an alternative version of this test (*Denver II*), contains more language items, new age scales, new categories of item interpretation and a behaviour rating scale. *Parents' Evaluation of Developmental Status (PEDS)*: Parents complete a 10 item questionnaire which is interpreted according to an algorithm. It is simple and quick, and unlike the Denver test, special items do not need purchasing. www.pedstest.com/ & 10958018

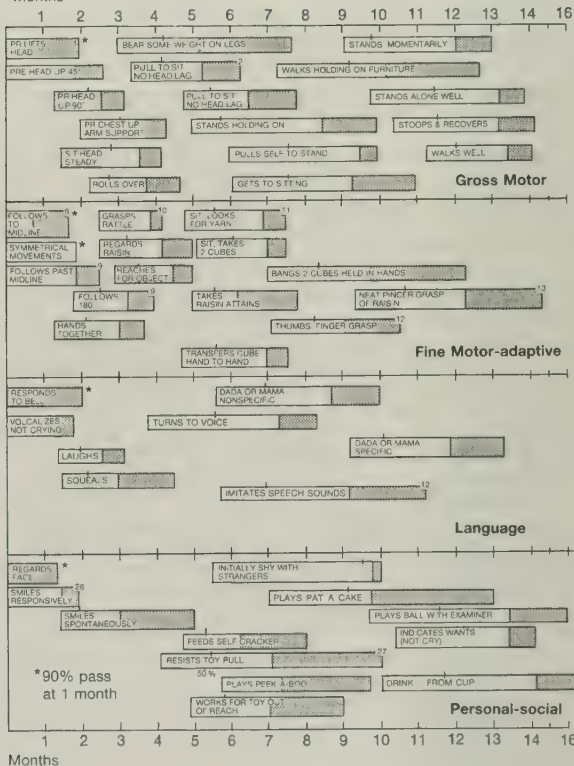
PR=Prone
SIT=Sit

Percent of children passing

25 50 75 90

TEST ITEM

Months



Percent of children passing

25 50 75 90

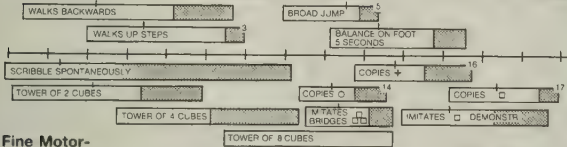
TEST ITEM

Months

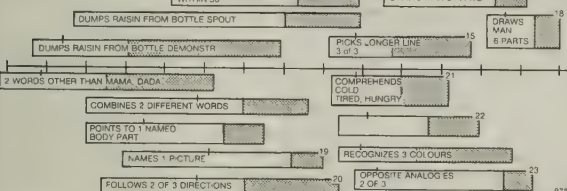
Years

12 13 14 16 18 20 22 24 2½ 3 3½ 4 4½ 5 6

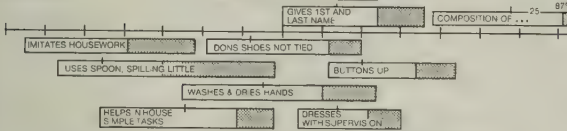
Gross Motor



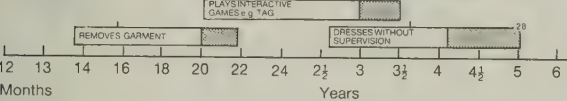
Fine Motor-adaptive



Language



Personal-social



Paediatric reference intervals

P = plasma; S = serum F = fasting

Biochemistry (1mmol = 1mEq/L)

Albumin ^P	36–48g/dL
Alk phos ^P	see below
(depends on age)	
α_1 -antitrypsin ^P	1.3–3.4g/dL
Ammonium ^P	2–25 μ mol/L; 3–35 μ g/dL
Amylase ^P	70–300u/L
Aspartate aminotransferase ^P	<40u/L
Bilirubin ^P	2–16 μ mol/L; 0.1–0.8mg/dL
Blood gases, arterial	pH 7.36–7.42
PaCO ₂	4.3–6.1 kPa; 32–46mmHg
PaO ₂	11.3–14.0 kPa; 85–105mmHg
Bicarbonate	21–25mmol/L
Base excess	–2 to +2mmol/L
Calcium ^P	2.25–2.75mmol/L; 9–11mg/dL
Neonates:	1.72–2.47; 6.9–9.9mg/dL
Chloride ^P	98–105mmol/L
Cholesterol ^{P,F}	\leq 5.7mmol/L; 100–200mg/dL
Creatine kinase ^P	< 80u/L
Creatinine ^P	25–115 μ mol/L; 0.3–1.3mg/dL
Glucose	2.5–5.3mmol/L; 45–95mg/dL
(lower in newborn. Fluoride tube)	
IgA ^S	0.8–4.5g/L (low at birth, rising to adult levels slowly)
IgG ^S	5–18g/L (high at birth, falls and then rises slowly to adult level)
IgM ^S	0.2–2.0g/L (low at birth, rises to adult level by one year)
IgE ^S	< 500u/mL

Iron ^S	9–36 μ mol/L; 50–200 μ g/dL
Lead ^{EDTA}	<1.75 μ mol/L; <36 μ g/dL
Mg ²⁺ P	0.6–1.0mmol/L
Osmolality ^P	275–295mosmol/L
Phenylalanine ^P	0.04–0.21mmol/L
Potassium ^P mean mmol/L:	Day 1: 6.4
	Day 2: 6; Day 3: 5.9 (later 4–5.5)
Protein ^P	63–81g/L; 6.3–8.1g/dL
Sodium ^P	136–145mmol/L
Transferrin ^S	2.5–4.5g/L
Triglyceride ^{F,S}	0.34–1.92mmol/L
	(\approx 30–170mg/dL)
Urate ^P	0.12–0.36mmol/L; 2–6mg/dL
Urea ^P	2.5–6.6mmol/L; 15–40mg/dL
Gamma-glutamyl transferase ^P	<20u/L

Hormones—a guide. ► Consult lab

Cortisol ^P	9am 200–700nmol/L
	midnight <140nmol/L (mean)
Dehydroepiandrosterone sulfate ^P :	
Day 5–11	0.8–2.8 μ mol/L (range)
5–11yrs	0.1–3.6 μ mol/L
17 α -Hydroxyprogesterone ^P :	
Days 5–11	1.6–7.5nmol/L (range)
4–15yrs	0.4–4.2nmol/L
T ₄ ^P	60–135nmol/L (not neonates)
TSH ^P	<5mu/L (higher on Day 1–4)

B=boy; EDTA=edetic acid; F=fasting
G=girl; P=plasma; S=serum.

Alk phos u/L: 0–½yr 150–600; ½–2yr 250–1000; 2–5yr 250–850; 6–7yr 250–1000; 8–9yr 250–750
10–11yr G=259–950, B \leq 730; 12–13yr G=200–750, B \leq 785; 14–15yr G=170–460, B=170–970;
16–18yr G=75–270, B=125–720; >18yr G=60–250, B=50–200.

Haematology mean \pm 1 s.d. Range $\times 10^9$ /L (median in brackets)

Day	Hb g/dL	MCV fl	MCHC%	Retic%	wcc	Neutrophils	Eosins	Lymphs	Monos
1	19.0 \pm 2	119 \pm 9	31.6 \pm 2	3.2 \pm 1	9–30	6–26 (11)	.02–.8	2–11	0.4–3.1
4	18.6 \pm 2	114 \pm 7	32.6 \pm 2	1.8 \pm 1	9–40				
5	17.6 \pm 1	114 \pm 9	30.9 \pm 2	1.2 \pm 2					

Weeks

1–2	17.3 \pm 2	112 \pm 19	32.1 \pm 3	0.5 \pm .03	5–21	1.5–10 (5)	.07–.1	2–17	0.3–2.7
2–3	15.6 \pm 3	111 \pm 8	33.9 \pm 2	0.8 \pm 0.6	6–15	1–9.5 (4)	.7–.1	2–17	0.2–2.4
4–5	12.7 \pm 2	101 \pm 8	34.9 \pm 2	0.9 \pm 0.8	6–15	(4)		(6)	
6–7	12.0 \pm 2	105 \pm 12	33.8 \pm 2	1.2 \pm 0.7	6–15	(4)		(6)	
8–9	10.7 \pm 1	93 \pm 12	34.1 \pm 2	1.8 \pm 1	6–15	(4)		(6)	

Months—all the following Hb values are Medians/Lower limit of normal

3	11.5/9	88/80			6–15	(3)		(6)	
6	11.5/9	77/70			6–15	(3)		(6)	
12	11.5/9	78/72			6–15	(3)		(5)	

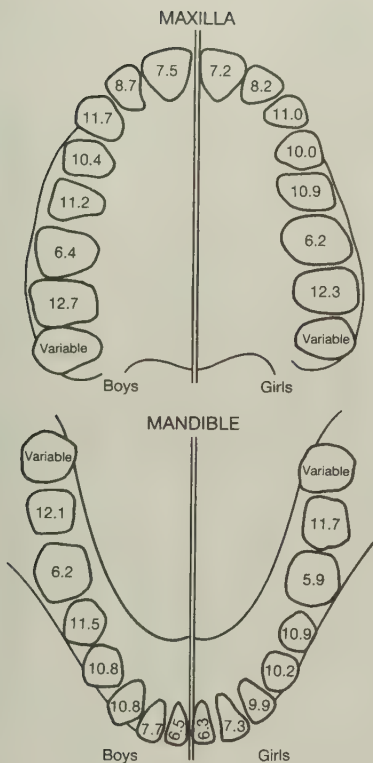
Years

2	11.5/9	78/74			6–15	(3)		(5)	
4	12.2/10	80/75			6–15	(4)		(4)	
6	13/10.4	82/75			5–15	(4.2)		(3.8)	
12	13.8/11	83/76			4–13	(4.9)		(3.1)	
14B	14.2/12	84/77			4–13	(5)		(3)	
14G	14/11.5								
16B	14.8/12	85/78	30–36	0.8–2	4–13	2–7.5 (5)	.04–.4	1.3–3.5	.2–.8
16G	14/11.5								
18B	15/13								

Note Basophil range: 0–0.1 $\times 10^9$ /L; B₁₂ \geq 150ng/L.

Red cell folate ^{EDTA} 100–640ng/mL. B=boys; G=girls.

Platelet counts do not vary with age; range: 150–400 $\times 10^9$ /L.



Mean times of eruption (in years) of the permanent teeth. ¹²

Deciduous teeth

	Months		Months
Lower central incisors	5-9	First molars	10-16
Upper central incisors	8-12	Canines	16-20
Upper lateral incisors	10-12	Second molars	20-30
Lower lateral incisors	12-15		

A 1-year-old has ~6 teeth; 1½ yrs ~12 teeth; 2 yrs ~16 teeth; 2¼ yrs ~20.

Centile tables

Age	Weight (kg)			Height (cm)			Skull circumference (cm)		
	Centile—Boys 3	50	97	3	50	97	3	50	97
Birth term	2.5	3.5	4.4	—	50	—	30	35	38
3 months	4.4	5.7	7.2	55	60	65	38	41	43
6 months	6.2	7.8	9.8	62	66	71	41	44	46
9 months	7.6	9.3	11.6	66	71	76	43	46	47
12 months	8.4	10.3	12.8	70	75	80	44	47	49
18 months	9.4	11.7	14.2	75	81	87	46	49	51
2 years	10.2	12.7	15.7	80	87	93	47	50	52
3 years	11.6	14.7	17.8	86	95	102	48	50	53
4 years	13	15	21	94	101	110			
5 years	14	19	23	100	108	117	49	51	54
6 years	16	21	27	105	114	124			
7 years	17	23	30	110	120	130			
8 years	19	25	34	115	126	137	50	52	55
9 years	21	28	39	120	132	143			
10 years	23	30	44	125	137	148			
11 years	25	34	50	129	142	154			
12 years	27	38	58	133	147	160	51	54	56
13 years	30	43	64	138	153	168			
14 years	33	49	71	144	160	176	53	56	58
15 years	39	55	76	152	167	182			
16 years	46	60	79	158	172	185			
17 years	49	62	80	162	174	187			
18 years	50	64	82	162	175	187			

Centile tables—Girls

Birth term	2.5	3.4	4.4	—	50	—	30	35	39
3 months	4.2	5.2	7.0	55	58	62	37	40	43
6 months	5.9	7.3	9.4	61	65	69	40	43	45
9 months	7.0	8.7	10.9	65	70	74	42	44	47
12 months	7.6	9.6	12.0	69	74	78	43	46	48
18 months	8.8	10.9	13.6	75	80	85	45	47	50
2 years	9.6	12.0	14.9	79	85	91	46	48	51
3 years	11.2	14.1	17.4	86	93	100	47	49	52
4 years	13	16	20	92	100	109			
5 years	15	18	23	98	107	116	48	50	53
6 years	16	20	27	104	114	123			
7 years	18	23	30	109	120	130			
8 years	19	25	35	114	125	136	50	52	54
9 years	21	28	40	120	130	142			
10 years	23	31	48	125	136	148			
11 years	25	35	56	130	143	155			
12 years	28	40	64	135	149	164	51	53	56
13 years	32	46	70	142	156	168			
14 years	37	51	73	148	160	172	52	54	57
15 years	42	54	74	150	162	173			
16 years	45	56	75	115	162	174			
18 years	46	57	75	—	—	—			

- Measure height exactly:**
- Feet together
 - Straight legs; loose arms
 - Buttocks, shoulders and heels touching wall
 - Hair firmly compressed
 - Ear canal level with infraorbital margin
 - Take reading in expiration.

More up-to-date charts are being designed to take into account new growth data, and to amalgamate American, who, and European centile divisions.¹

¹ The 1990 nine centile UK charts are available from Harlow Publishing, Maxwell St, South Shields, NE33 4PU Tel. 0191 427 0195. See TJ Cole 1994 *BMJ* i 641 & DMB Hall 1995 *BMJ* ii 583. **NB:** the 1995 Castlemead (Buckler) charts have not met with approval from paediatric growth experts because the upper centile on the weight chart is too low, classifying too many children as obese—but these charts have the (unproven) advantage of being based on serial measurements, not cross-sectional data.

BOYS BMI CHART

(BIRTH - 20 YEARS): United Kingdom cross-sectional reference: 2002/1

Name
 D.O.B. [DDMMYY] / /
 NHS No.

Body Mass Index (BMI)

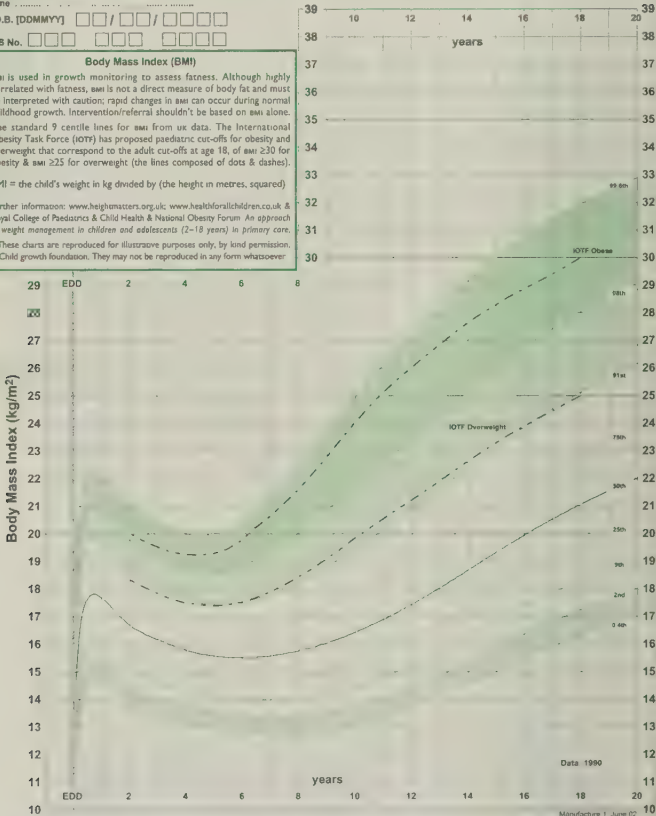
BMI is used in growth monitoring to assess fatness. Although highly correlated with fatness, BMI is not a direct measure of body fat and must be interpreted with caution; rapid changes in BMI can occur during normal childhood growth. Intervention/referral shouldn't be based on BMI alone.

The standard 9 centile lines for BMI from UK data. The International Obesity Task Force (IOTF) has proposed paediatric cut-offs for obesity and overweight that correspond to the adult cut-offs at age 18, of BMI ≥ 30 for obesity & BMI ≥ 25 for overweight (the lines composed of dots & dashes).

BMI = the child's weight in kg divided by (the height in metres, squared)

Further information: www.heightmatters.org.uk; www.healthforallchildren.co.uk & Royal College of Paediatrics & Child Health & National Obesity Forum 'An approach to weight management in children and adolescents (2-18 years) in primary care'.

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Reference

Body Mass Index reference curves for the UK, 1990 (TJ Cole, JV Freeman, MA Preece) *Arch Dis Child* 1995; 73: 25-29

Establishing a standard definition for child overweight and obesity: international survey, (Cole TJ, Bellizzi MC, Flegal KM, Dietz WH) *BMJ* 2000; 320: 1-6



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GIRLS BMI CHART

(BIRTH - 20 YEARS): United Kingdom cross-sectional reference: 2002/1

Name

D.O.B. [DDMMYY] / /

NHS No.

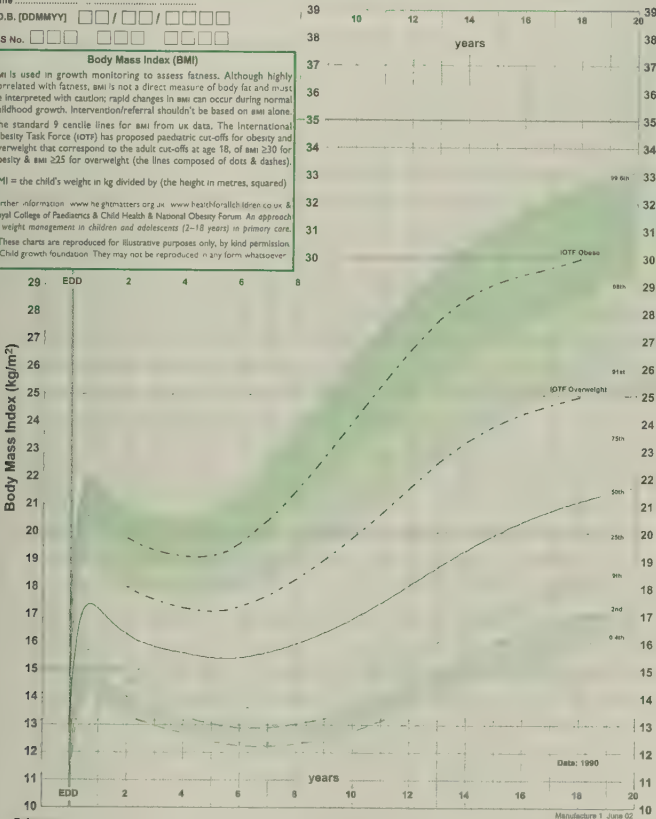
Body Mass Index (BMI)

BMI is used in growth monitoring to assess fatness. Although highly correlated with fatness, BMI is not a direct measure of body fat and must be interpreted with caution; rapid changes in BMI can occur during normal childhood growth. Intervention/referral shouldn't be based on BMI alone. The standard 9 centile lines for BMI from UK data. The International Obesity Task Force (IOTF) has proposed paediatric cut-offs for obesity and overweight that correspond to the adult cut-offs at age 18, of BMI ≥ 30 for obesity & BMI ≥ 25 for overweight (the lines composed of dots & dashes).

BMI = the child's weight in kg divided by (the height in metres, squared)

Further information: www.heightmatters.org.uk, www.healthforallchildren.co.uk & Royal College of Paediatrics & Child Health & National Obesity Forum. An approach to weight management in children and adolescents (2-18 years) in primary care.

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












































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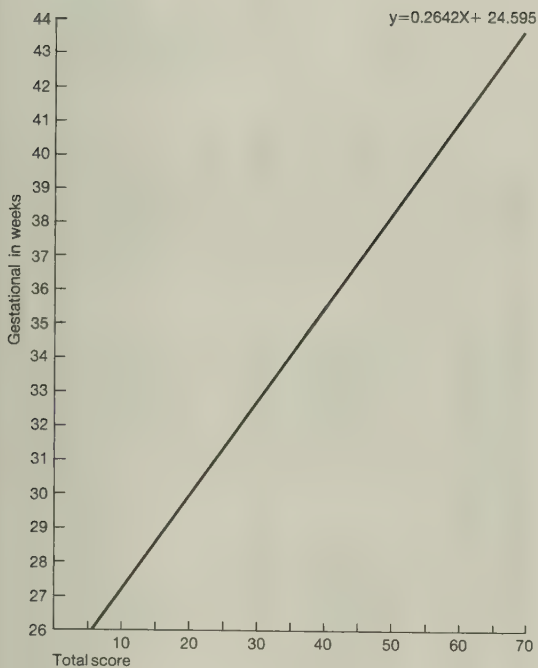


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Assessment of gestational age: Dubowitz system

NEURO-LOGICAL SIGN	SCORE					
	0	1	2	3	4	5
POSTURE						
SQUARE WINDOW	 90°	 60°	 45°	 30°	 0°	
ANKLE DORSI-FLEXION	90° 	75° 	45° 	20° 	0° 	
ARM RECOIL	180° 	90-180° 	<90° 			
LEG RECOIL	180° 	90-180° 	<90° 			
POPLITEAL ANGLE	180° 	160° 	130° 	110° 	90° 	<90° 
HEEL TO EAR						
SCARF SIGN						
HEAD LAG						
VENTRAL SUSPENSION						

Neurological criteria. (By kind permission of the *Journal of Paediatrics*.)



Assessment of gestational age: Dubowitz system. Graph for reading gestational age from total score. (By kind permission of the *Journal of Paediatrics*.)

Assessment of gestational age: Dubowitz system

Physical (external) criteria (for neurological criteria, see p 298)

External sign	Score				
	0	1	2	3	4
Oedema	Obvious oedema hands and feet: pitting over tibia	No obvious oedema hands and feet: pitting over tibia	No oedema		
Skin texture	Very thin, gelatinous	Thin and smooth	Smooth: medium thickness Rash or superficial peeling	Slight thickening Superficial cracking and peeling, especially hands and feet	Thick and parchment-like: superficial or deep cracking
Skin colour (infant not crying)	Dark red	Uniformly pink	Pale pink: variable over body	Pale Only pink over ears, lips, palms, or soles	

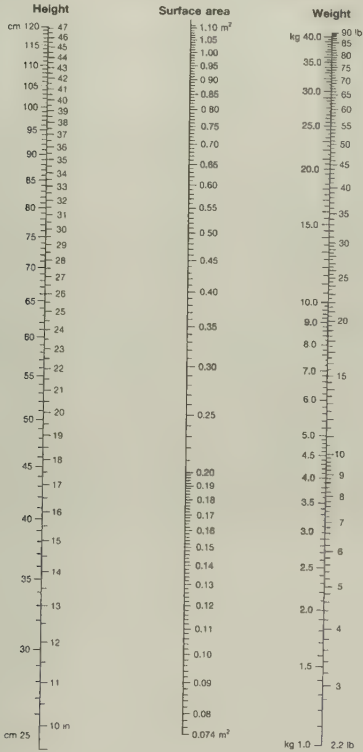
Skin opacity (trunk)	Numerous veins and venules clearly seen, especially over abdomen	Veins and tributaries seen	A few large vessels clearly seen over abdomen	A few large vessels seen indistinctly over abdomen	No blood vessels seen
Lanugo (over back)	No lanugo	Abundant long and thick over whole back	Hair thinning especially over lower back	Small amount of lanugo and bald areas	At least half of back devoid of lanugo
Plantar creases	No skin creases	Faint red marks over anterior half of sole	Definite red marks over more than anterior half; indentations over less than anterior third	Indentations over more than anterior third	Definite deep indentations over more than anterior third
Nipple formation	Nipple barely visible: no areola	Nipple well defined: areola smooth and flat diameter <0.75cm	Areola stippled, edge not raised: diameter <0.75cm	Areola stippled, edge raised diameter >0.75cm	
Breast size	No breast tissue palpable	Breast tissue on one or both sides <0.5cm diameter	Breast tissue both sides: one or both 0.5—1.0cm	Breast tissue both sides: one or both >1cm	

Assessment of gestational age: Dubowitz system

Physical (external) criteria (for neurological criteria, see p 298)

External sign	Score				
	0	1	2	3	4
Ear form	Pinna flat and shapeless, little or no incurving of edge	Incurving of part of edge of pinna	Partial incurving whole of upper pinna	Well-defined incurving whole of upper pinna	
Ear firmness	Pinna soft, easily folded, no recoil	Pinna, soft, easily folded, slow recoil places, ready recoil	Cartilage to edge of pinna, but soft in places, ready recoil	Pinna firm, cartilage to edge, instant recoil	
Genitalia					
●Male	Neither testis in scrotum	At least one testis high in scrotum	At least one testis right down		
●Female (with hips half abducted)	Labia majora widely separated, labia minora protruding	Labia majora almost cover labia minora	Labia majora completely cover labia minora		

Nomogram for calculating the body surface area of children



Reference intervals:hypertension

Age (Years)	Significant hypertension		Severe hypertension	
	Systolic	Diastolic	Systolic	Diastolic
At birth	≥96mmHg*	≥106mmHg*		
<2	≥112mmHg	≥74mmHg ^{k4}	≥118mmHg	≥82mmHg ^{k4}
3–5	≥116mmHg	≥76mmHg ^{k4}	≥124mmHg	≥84mmHg ^{k4}
6–9	≥122mmHg	≥78mmHg ^{k4}	≥130mmHg	≥86mmHg ^{k4}
10–12	≥126mmHg	≥82mmHg ^{k4}	≥134mmHg	≥90mmHg ^{k4}
13–15	≥136mmHg	≥86mmHg ^{k5}	≥144mmHg	≥92mmHg ^{k5}
16–18	≥142mmHg	≥92mmHg ^{k5}	≥150mmHg	≥98mmHg ^{k5}

Take ≥3 BPs (snugly fitted cuff of bladder-width >75% of upper arm length) >1 week apart (in general) before diagnosing hypertension. As the 5th Korotkoff sound (^{k5}) is inaudible, use ^{k4} for diastolic BPs, until adolescence.⁶ Some BP standards break populations by age, sex, and height, but this doesn't greatly change hypertension prevalence. ²⁰⁰¹ medline:11368459 &: 3797155

Ambulatory BP recording is helpful, eg showing that white coat hypertension is about as common in children as in adults. ²⁰⁰¹ medline:11317054 &: 10863171

*Doppler may be needed—it is often only possible to record systolic BPs

Reference intervals may need adjusting if the child is taller or shorter than expected for age (tables are available); compared with a 50th height-centile child the reference interval is 1–4mmHg lower for a child on the 5th height-centile (1–4mmHg higher if on 95th height-centile). medline:8885941

Causes in neonates: Renal artery stenosis (or thrombosis), congenital renal malformations, coarctation of the aorta, bronchopulmonary dysplasia.

Infants: Renal parenchymal diseases, aorta coarctation, renal artery stenosis.

6–10 years old: Renal artery stenosis, renal diseases, primary hypertension.

Adolescence: Primary hypertension, renal parenchymal diseases.

Reference intervals: nutritional requirements for preterm babies

Water see p 251 An intake of 180mL/kg/day (range 150–200mL) of human or formula milk meets the water needs of very low birth weight infants (VLBW; <1500g) under normal circumstances. In infants with heart failure water restriction is necessary (eg 130mL/kg/day).

Energy 130kcal/kg/day (range 110–165) meets the needs of the LBW infant in normal circumstances, and can be provided by formulas with similar energy density to human milk (65–70kcal/dL) in a volume of 180–200mL/kg/day. If a higher energy density is required keep it <85kcal/dL. The problem with energy densities above this is fat lactobezoars and u&e imbalance.

Protein Aim for between 2.25g/100kcal (2.9g/kg/day when fed at 130kcal/kg) and 3.1g/100kcal (4g/kg/day). Lysine should be as high as possible. Precise guidelines on taurine and whey:casein ratios cannot be given. At present LBW formulas are whey-predominant. Signs of protein deficiency: a low plasma urea and prealbumin.

Fat Aim for 4.7–9g/kg (fat density 3.6–7.0g/100kcal). Longer chain unsaturated fatty acids (>C12) are better absorbed than saturated fatty acids. Aim to have ≥4.5% of total calories as the essential fatty acid linoleic acid (500mg/100kcal).

www.maabarot-export.co.il/baby/milk/stage2-2.htm

Carbohydrates Aim for 7–14g/100kcal (<11g/100dL), with lactose contributing 3.2–12g/100kcal (<11g/100dL). Lactose is not essential; substitutes are glucose (but high osmolality may cause diarrhoea) or sucrose (± starch hydrolysates, eg corn syrup oils).

Vitamins See p 247 & ESPGAN committee.

medline:8793820

Elements Na⁺: 6.5–15mmol/L. K⁺: 15–25.5mmol/L. Ca²⁺ 1.75–3.5mmol/100kcal. PO₄³⁻: 1.6–2.9mmol/100kcal. Ca²⁺:PO₄³⁻ ratio: 1.4–2.0:1. Magnesium: 0.25–0.5mmol/100kcal. Iron: if breast fed, give 2–2.5mg Fe/kg/day (the recommended total intake). Formula-fed infants may need a supplement to achieve this. Iodine: 10–45µg/100kcal. Manganese: 2.1µmol/100kcal. ►1cal=4.18 joules.

APLS procedures—intraosseous transfusion

(APLS stands for advanced paediatric life support.)

Immediate vascular access is required in paediatric and neonatal practice in the following circumstances: cardiopulmonary arrest, severe burns, prolonged status epilepticus, hypovolaemic and septic shock. In many cases rapid intravenous access is not easily obtained, and intraosseous infusion is a relatively safe, easy, and effective means of obtaining vascular access, and is recommended for life-threatening paediatric emergencies in which other methods of access have failed.

medline:109691

Contra-indications Osteoporosis, osteogenesis imperfecta, and infection or fracture at the site of insertion.

Technical aspects *Learn from an expert.* The following is one technique

Preparation: Set trolley: Dressing pack, Betadine®, needles, 10mL syringe, lidocaine (=lignocaine) 1% (5mL), scalpel, intraosseous needle, paediatric infusion set, 10mL 0.9% saline, adhesive tape.

Choosing the site of insertion: The proximal tibia is the best site. Other sites are the distal tibia, or distal femur. Choose a point in the midline on the flat anteromedial surface of the tibia, 2 finger-breadths below the tibial tuberosity. The patient's leg should be restrained, with a small support placed behind the knee.

Procedure:

- Sterilize the skin with antiseptic, infiltrate with lidocaine as necessary (Puncturing the skin with the scalpel is not usually necessary.)
- Insert the intraosseous needle at an angle of 60–90 degrees away from the growth plate, and advance with a boring or screwing motion into the marrow cavity. Correct location of the needle is signified by a decrease in resistance on entering the marrow cavity.
- Stabilize the needle and verify the position by aspirating marrow, or by the easy flushing of 5–10mL of 0.9% saline, without any infiltration of surrounding tissue. The needle should stand upright without support but should be secured with tape.
- Flushing with heparinized saline (0.9%) may prevent clotting.
- Connect to iv infusion—but better flow rates are often achieved by syringing in boluses of fluid (standard bolus is 20mL/kg of crystalloid or colloid).

Complications These are infrequent, but there may be extravasation of fluid, or cellulitis, fractures, osteomyelitis, pain, and fat or bone microemboli. These are more common with prolonged use—so intraosseous infusion should be discontinued as soon as conventional iv access is attained.

NB: Intraosseous delivery may also be used in adults.

medline:10658988

►► Anaphylaxis

Adrenaline = epinephrine)

Doses of *IM* adrenaline ►Note the ampoule strength! 1:1000 or 1:10000.

Age	mL of 1:10,000*	Age	mL of 1:1000
3–5 months	0.5mL	2yrs**	0.2mL
6–11 months	0.75mL	3–4yrs**	0.3mL
12 months	1.0mL	5yrs**	0.4mL
		6–12yrs**	0.5mL; adult 0.5–1mL

Doses may need repeating every 15min, until improvement occurs. Also:

- O₂ (± IPPV) • Hydrocortisone (4mg/kg iv) • Chlorpheniramine (0.2mg/kg)
- Colloid (20mL/kg ivi). **NB:** weight (kg) ≈ 2(age in yrs + 4). ok if 1–10yrs.

*This is 0.01% weight/volume, ie 1mg in 10mL.

**For underweight children, halve the dose, or, if weight known, give 10µg/kg (=0.01mL/kg of 1:1000). Epinephrine dose by endotracheal tube: 100µg/kg. If bronchospasm is a feature, give salbutamol 2mg nebulized too. After the emergency, take blood at 1–5h for mast cell tryptase; refer to an allergist. Self-use of pre-loaded pen injections may be needed (eg EpiPen®).

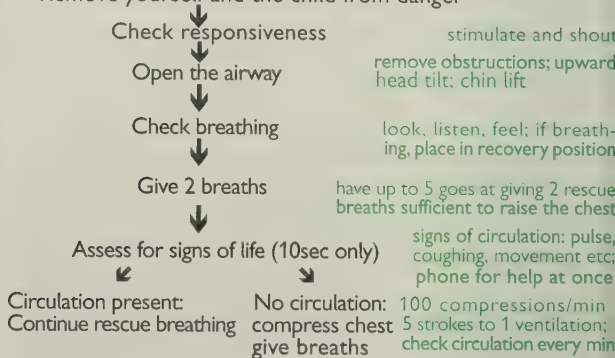
medline~9572760

EpiPen® contains 0.3mg of adrenaline. This is suitable if weight is more than 30kg. If 15–30kg, use Anapen Junior® or EpiPen Junior®, which both contain 0.15mg of adrenaline.

APLS/European paediatric basic life-support algorithm*

This algorithm assumes no equipment and that only one rescuer is present

Remove yourself and the child from danger



Compress sternum to $\frac{1}{3}$ of the chest's depth; use the heel of one hand (or, in babies, with both your thumbs, with your hands encircling the thorax). If >8yrs, the adult 2-handed method is OK. For an infant, 2 fingers are sufficient, positioned in the middle of a line joining the nipples. Perform resuscitation for ~1 min before going for help. Remove the cause, if possible. Causes are: drowning; PE; trauma; electrocution; shock; hypoxia; hypercapnia; hypothermia; U&E imbalance; drugs/toxins, eg adrenaline (epinephrine), digoxin, and blue-ringed octopi. medline:3573123

How to give the rescue breaths to a child Ensure head tilt and chin lift. Pinch the soft part of his nose. Open his mouth a little, but maintain chin up. Take a breath, and place your lips around his mouth (good seal). Blow steadily into his mouth over 1–1.5sec. Does his chest rise? Take your mouth away, and watch for the chest to fall. Take another breath, and repeat this sequence up to 5 times. **With an infant** Do as above, but cover the nasal apertures and the mouth with your lips. If the chest does not move, respiratory obstruction may exist ►►move on to 'Removing foreign body' sequence for obstructed airway—ie:

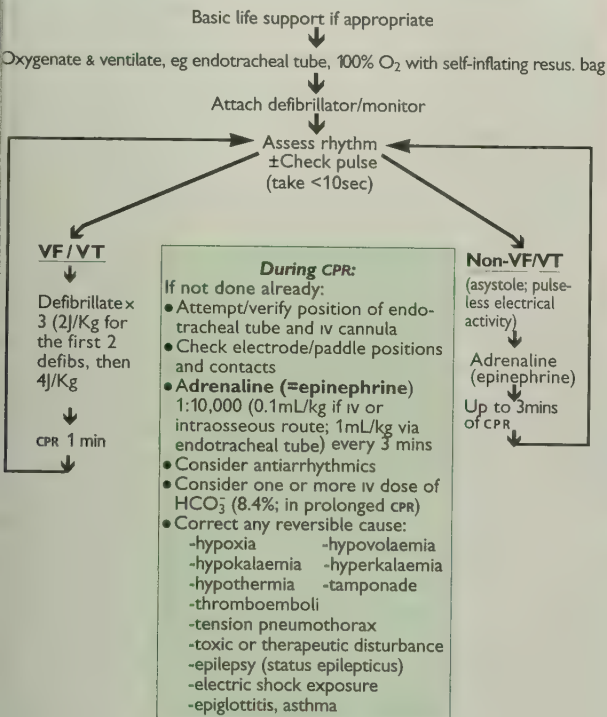
- Remove any obvious obstructions. Recheck that there is adequate head tilt and chin lift, but do not overextend the neck.
- Do up to 5 back blows between the scapulae to dislodge hidden obstructions (hold prone, positioning the head lower than chest).
- If this fails, do 5 chest thrusts: turn to supine; over 12sec, give 5 sternal thrusts (same position as for compressions, but be sharper and more vigorous). Remove any foreign bodies which have become visible.
- Tilt head upwards; lift chin to reopen the airway, and assess breathing.
- If not breathing; do 5 more rescue breaths: does the chest move now?
- If not, for a child, give 5 abdominal thrusts (directed towards diaphragm); use the upright position if the child is conscious; supine if not.
- Repeat these sequences until breathing is OK, alternating chest and abdominal thrusts. *Do not give abdominal thrusts to infants.*

When breathing place in the recovery position—as near to the true lateral position as possible, with mouth dependent to aid draining of secretions. The position must be stable (eg use pillows placed behind back). The degree of movement is determined by risk of spinal injury.

*www.resus.org.uk/SiteIndex.htm#medical & www.ncems.org/pdf/emsc/manual.pdf

Cardiac arrest: paediatric advanced life-support¹

Each step assumes the previous one has been unsuccessful



Treat acidosis with good ventilation. Sodium bicarbonate may worsen intracellular acidosis and precipitate arrhythmias, so use it only in severe acidosis (eg 1mmol/kg, by slow iv bolus). NB: in adults bicarbonate is normally recommended only after prolonged resuscitation. In children, particularly neonates, the heart is often the last thing to stop—so when it does, severe acidosis may *already* be present.

4. Psychiatry

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Other topics

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Sources: This is our best-sourced Chapter 4 ever, using *BMJ*, Cochrane, *EBM*, etc., but note that 'too great an emphasis on *EBM* oversimplifies the complex and interpersonal nature of care'. In psychiatry it is better to think of each encounter with a patient as an opportunity for creative thought and intimate dialogue, not as an opportunity to apply a previously agreed set of rules. medline.11772844

Relevant pages elsewhere: Puerperal psychosis, p 148; consultations, p 422–4; social/ethnic matters, p 462 & 436; prescribing & compliance, p 460; children, p 206.

This chapter should be read with the *Diagnostic and Statistical Manual of Mental Disorders* (4th edn). But note that this publication is really for research purposes, and its definitions may be too rigid and rigorous for general psychiatric purposes—ie its specificity is excellent, but its sensitivity is questionable.

Affective (mood) disorders:

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No system can be equally successful in guiding treatment **and** in illustrating basic biological distinctions. For example, depression spans the neurosis/psychosis division, and mania has both affective (mood) and psychotic components.

- There is no such thing as a correct classification, only more or less useful ones. Bear in mind that in ancient times (as reported by Borges) one approved classification of animals had as its first 6 categories: 1 Animals that belong to the Emperor. 2 Embalmed ones. 3 Those that are trained. 4 Suckling pigs. 5 Mermaids. 6 Fabulous ones. We do not know how many mermaids and other fabulous beings are included in the above classificatory system.
- Not all the diagnostic categories have the same value. If a patient with features of schizophrenia (eg hallucinations) is later found to have an organic psychosis (eg from a brain tumour) the schizophrenic element is usually subsumed under the more powerful diagnostic category of organic illness. This is an example of the 'hierarchy of diagnosis', in which organic illness 'trumps' schizophrenia and affective disorders, which in turn trump neurosis, but this does preclude multiple diagnoses, eg schizophrenia with depression and substance abuse disorder.
- No one is just a member of a class, and it is a common error to abandon patients, once they have been categorized. Because of its tragic essence, it is possible for mental illness to be a humanizing experience for those affected (patient, family, and friends)—as well as being an intensely destructive one, and there is much to be said for the idea that we should embrace our patients, rather than analyse them. But being holistic must, on occasion, give ground to reductionism, for diagnosis is the single best tool we have to help our patients.

Introduction to psychiatry

Doctors have never been very important because nothing that happens to our bodies ever really matters. In historical terms, the importance of what happens to our bodies rarely outlives our own times. Even the exceptions to this prove a different rule: for example, we recall forever the human images burnt on to stone at their moment of immolation in Nagasaki and Hiroshima. Surely what happened to them was important, and transcended the times in which they lived? True. But it is all the more true that it isn't the way they lived, but the way they died that is important, and it is not the body nor its image on stone which is important, but the image left on our minds. And so it is with psychiatrists, psychologists, and psychiatric nurses who play such a rôle in determining whether we live and die in hope or despair, who mitigate our madness giving meaning, purpose, and dignity to the mental sufferings of so many of us. So when we think of these men and women going about their work, we think of them burnishing humanizing images not on stone, but on the mind itself. How they do it is the subject of this chapter.

Mental health and mental illness

What is mental health? Ideally, healthy humans have:

- An ability to love and be loved. Without this cardinal asset, human beings, more than all other mammals, fail to thrive.
- The power to embrace change and uncertainty without fear—and to face fear rationally and in a spirit of practical optimism.
- A gift for risk-taking free from endless worst-case-scenario-gazing.
- Stores of spontaneous *joie de vivre*, and a wide range of emotional responses (including negative emotions, such as anger; these may be important for motivation, as well as being a natural antidote to pain).
- Efficient contact with reality: not too little; not too much. (As T S Eliot said, humankind cannot bear very much reality.)
- A rich fantasy world enabling hope and creativity to flourish.
- A degree of self-knowledge to encourage the humane exercising of the skill of repairing the self and others following harm.
- The strength to say 'I am wrong', and to learn from experience.
- An adequate feeling of security and status within society.
- The ability to satisfy the requirements of the group, combined with a freedom to choose whether to exercise this ability.
- Freedom of self-expression in whatever way he or she wants.
- The ability to risk enchantment and to feel a sense of awe.
- The ability to gratify his own and others' bodily desires.
- A sense of humour to compensate should the foregoing be unavailable.

Happiness need not be an ingredient of mental health; indeed the merely happy may be supremely vulnerable. All that is needed is for their happiness to be removed. The above are important as they are what a person needs should this misfortune befall him. The above may also be seen as a sort of blueprint for our species' survival.

The sense of mental illness Whenever a person's abnormal thoughts, feelings, or sensory impressions cause him objective or subjective harm that is more than transitory, a mental illness may be said to be present. Very often the harm is to society, but this should not be part of the definition of mental illness, as to include it would open the door to saying that, for example, all rapists or all those opposing the society's aims are mentally ill. If a person is manic, and not complaining of anything, this only becomes a mental illness (on the above definition) if it causes him harm as judged by his peers (in the widest sense of the term). One feature of mental illness is that one cannot always rely on the patient's judgment, and one has to bring in the judgment of others, eg family, GP, or psychiatrist. If there is disparity of judgment, there is much to be said for adopting the principle of 'one person one vote', provided it can be shown that the voters are acting solely in the interests of the person concerned. The psychiatrist has no special voting rights here—otherwise, the concepts of mental health and illness become dangerously medicalized. Just because the psychiatrist or GP is not allowed more than one vote, this does not stop them from transilluminating the debate by virtue of their special knowledge.

For convenience, English law saves others from the bother of specifying who has a mental illness by authorizing doctors to act for them. This is a healthy state of affairs only in so far as doctors remember that they have only a small duty to society, but a larger duty to their patient (not, we grant, an overriding duty in all instances—eg when murder is in prospect).

Learning Disabilities or Mental Retardation This is a condition of arrested or incomplete development of mind which is especially characterized by subnormality of intelligence.

Dignity on psychiatric wards

Apart from showing sympathy, nothing is more to be desired than giving our patients dignity—not the dignity that they deserve (who among us would merit an ounce of dignity on this measure?) but the dignity that confirms to our patients that, mad, bad, or rambling, they are, root and branch, body and mind, just as human as their doctors—perhaps more so, because they are suffering.* *Let patients:*

- Decide on mode of address, 'Miss Hudson' may be preferred to 'Agatha, dear'. Dignity entails giving choices, and then respecting them.
- Know who we are (eg wear name badges).
- Wear their own clothes—and clothe them decently if they have none.
- Choose whether to take part in research—and whether to see students.
- Have personal space—both to stow their belongings, and to walk in, in private, whether alone, or with visitors.
- Participate in their treatment plans. Explain about common side-effects.
- Know what to do if a crisis develops. It's a great help to know that you will be seen in 4h rather than be left to moulder all weekend.

These 7 points are the basis of the UK *patient's charter* in mental health, and they are also congruent with the UK's Human Rights Act (1998).¹ This Act lays out the right to life, freedom from inhuman or degrading treatment or torture, respect for privacy, the right to a fair hearing and freedom of expression. Some of these rights are inalienable, and unrevokable, even in time of war. So defences such as 'the ward was very busy on that day' or 'there was no money to purchase this service' are unlikely to impress judges.

Our patients' other mental health hierarchy?

Maslow states that a healthy personality (ie fully functioning and self-actualizing) entails the meeting of a hierarchy of needs:

- 1 Biological needs (eg oxygen, food, water, warmth).
- 2 Safety needs (no present threats to safety).
- 3 Love, affection, and 'belongingness' needs.
- 4 Esteem needs (self-respect, and need for respect from others: see *above*).
- 5 Self-actualization needs (the need to *be* and *do* that which we feel born to do—his or her calling). If these needs are not met, restlessness and tenseness result. To meet this need we might teach ourselves and others:
 - To be aware of the inner self and to understand our inner nature.
 - To transcend cultural conditioning.
 - To transcend the trifling and to grapple with life's serious problems.
 - To refresh consciousness by appreciating beauty and all good things.
 - To feel joy and the worth of living.

www.connect.net/georgen/maslow.htm

¹ Violation of rights to a fair hearing and to privacy can be defended on grounds of public interest, protection of health or morals and, perhaps, economics. See www.coe.fr/eng/legaltxt/e-dh.htm

Do our sufferings make us more human? Only if we can breath meaning into them. Suffering does not make us more human because it has no meaning; but there is a kind of offering 'which is a more effective key, a more rewarding principle for exploring the world than thought and action than personal good fortune'. [Deitrich Bonhoeffer]. This suffering makes our souls. In psychiatric practice, not all our patients regret their illnesses: sometimes, in retrospect, these patients refer to their *breakthrough*, not their *breakdown*. It is this power to grow and to transform experience which is human and humanizing.

Odd ideas

It is important to decide whether a patient has delusions, hallucinations, or a major thought disorder (see below), because if these are present the diagnosis must be: schizophrenia, affective disorder, an organic disorder, or a paranoid state—and it is *not* a neurosis or a personality disorder.

Patients may be reluctant to reveal odd ideas. Ask gently: 'Have you ever had any thoughts which now seem odd; perhaps that there is a conspiracy against you, or that you are controlled by outside voices or the radio?'

Hallucinations are auditory, visual, gustatory, or tactile sensations experienced in the absence of a stimulus. Often-encountered pre- or post-sleep (hypnagogic/hypnopompic) hallucinations don't indicate pathology. A pseudo-hallucination is one in which the patient knows the stimulus is in the mind (eg a voice heard within him- or herself, rather than over the left shoulder). They are more common, and needn't indicate mental illness, but they may be a sign that a genuine hallucination is being internalized before being disbanded (ie the person hears a voice but no longer believes it represents an outside agent). Tactile or visual hallucinations (without auditory hallucinations) suggest an organic disorder (eg alcohol withdrawal, or Charles Bonnet syndrome, p 500). 2–4% of the general population experience auditory hallucinations, but only ~30% of these have a mental illness.

Delusions are beliefs which are held unshakably, irrespective of counter-argument, and which are unexpected, given the patient's cultural background. If the belief arrives fully formed, and with no antecedent events or experiences to account for it, it is said to be *primary*, and is suggestive of schizophrenia (or genius). Such delusions form around a 'delusional perception', as illustrated by the patient who, on seeing the traffic lights go green (the delusional perception) knew that he had been sent to rid his home town of materialism. Careful history-taking will reveal that delusions are often *secondary*—for example, a person who is psychotically depressed may come to think of himself as being literally worthless.

Obsessional ideas The patient cannot help feeling that other people are taking notice of the very thing he feels ashamed of. What distinguishes this from a delusion is that he knows the thoughts come from within himself, and are excessive. It is important to distinguish delusions and hallucinations from obsessional thoughts. With any odd idea, get a clear description:

- Hearing the thought as a voice (a hallucination, eg from a psychosis).
- The voice is 'put into my head'—thought insertion (hallucination + delusion).
- The voice is 'my own voice' but intrusively persistent (obsessional neurosis).

Flight of ideas This is a bizarre or incongruent transition from one idea to another. (*Mania*—flight of ideas, p 354; *schizophrenia*, p 358).

If hallucinations or delusions are present, ask yourself: 1 What other evidence there is of psychopathology. Hearing the voice of one's dead spouse is common, and does not indicate mental illness. 2 Could the odd ideas be adaptive—and the patient be better off 'ill'? A woman once believed she saw planes flying over her home, and that this data was taken from her head by the British Ministry of Defence. She 'knew' she was playing a key rôle in the defence of Britain. When she was cured of these delusions she committed suicide. An odd story—or is it so odd? According to the great poets, everything we cherish is an illusion, especially our sense of distinctive self-hood, and without this primordial delusion, madness beckons.

Some causes of odd ideas

A typical problem is trying to diagnose a young man presenting with hallucinations and or delusions. The question often is: *Are these odd ideas due to schizophrenia, drug abuse, or physical illness?*

- Most auditory hallucinations not associated with falling asleep or waking up are caused by schizophrenia or depression.
- In 90% of those with non-auditory hallucinations (eg seeing things), the cause is substance abuse, drug withdrawal, or physical disease.
- Evidence that substance abuse is to blame includes:
 - *The history*: Ask the patient, the family, and friends about abuse. Be precise about timing. If ≥ 4 weeks elapse between abuse and starting of odd ideas, substance abuse is an unlikely cause.
 - *Severity of symptoms*: If symptoms are severe, and the quantity of drug ingested is trivial, the drug is unlikely to be causative.
 - *Drug-seeking behaviour*: Be on the lookout for this.
 - *Physical examination*: This may reveal signs of drug abuse (eg injection marks \pm cellulitis), chronic alcohol abuse (eg spider naevi, liver palms, atrophic testes), or a physical medical illness (eg brain tumour).
 - *Blood or urine tests* may disclose the substance abused or give a hint of abuse (mcv \uparrow and gamma gt \uparrow in alcohol abuse).
 - *Imaging*: Consider CNS imaging if the patient is elderly with nothing to suggest substance abuse, or if there are CNS signs.
- NB: middle-age is not a typical time for schizophrenia to present: alcohol abuse or a primary CNS condition is more likely.
- Diagnosing a *substance-induced psychotic disorder* implies that the patient responds to the hallucinations or delusions as if they were real. If the patient recognizes the hallucinatory nature of the experience, then consider diagnosing *substance intoxication*, *substance withdrawal* or, if there is past but no current exposure to hallucinogens, the diagnosis may be 'flashbacks'—ie *hallucinogen persisting perception disorder*. This condition presents episodically up to 5yrs after exposure to an hallucinogen, with flashback hallucinations, any visual hallucinations or phenomenon such as: geometric visual hallucinations, seeing flashes of colour, or intensified colours, seeing dots, spots, or flashes, seeing trailing images or after-images, seeing complementary coloured images of objects gone from view, seeing halos, seeing things too small (micropsia), or seeing things too big (macropsia). These phenomena may be self-induced or triggered by darkness, stress, or fatigue. (DSM-IV 313)

“We entered an omnibus to go someplace...at the moment I put my foot on the step the idea came to me without anything in my former thoughts seeming to have paved the way for it, that the transformations I had used to define the Fuchsian functions were identical with those of non-Euclidean geometry...the idea came to me with...brevity, suddenness, and immediate certainty.” A Koestler *The Act of Creation* 115

Introduction to psychiatric skills

Only people can change people: 'Alyosha's arrival seemed to have a sobering effect on him. . . as though something had awakened in this prematurely aged man that had long been smothered in his soul . . . "For you see, my dear boy, I feel that you are the only man in the world who has not condemned me"'. ' (Father speaking to his son in *The Brothers Karamazov*, 25, Dostoyevsky.)

On beginning psychiatry you are likely to feel unskilled. A 'medical' problem will come as a relief—you know what to do. Do not be discouraged: you already have plenty of skills (which you will take for granted). The aim of this chapter is to build on these. No one can live in the world very long without observing or feeling mood swings, and without devising ways to minimize what is uncomfortable, and maximize what is desirable. Anyone who has ever sat an important exam knows what anxiety is like, and anyone who has ever passed one knows how to master anxiety, at least to some extent. We have all survived periods of being 'down', and it is interesting to ask how we have done this. The first element is time. Simply waiting for time to go by is an important psychotherapeutic principle. Much of what passes as successful treatment in psychiatry is little more than protecting the patient, while time goes by, and his (and his family's) natural processes of recovery and regeneration bring about improvement. Of course, there are instances when, to wait for time to go by could lead to fatal consequences. But this does not prevent the principle from being a useful one.

Another skill with which we are all more or less adept is *listening*. One of the central tenets of psychiatry is that it helps our patients just to be listened to. Just as we all are helped by talking and sharing our problems, so this may in itself be of immense help to our patients, especially if they have been isolated, and feel alone—which is a very common experience.

Just as spontaneous regeneration and improvement are common occurrences in psychiatry, so is relapse. Looking through the admissions register of any acute psychiatric ward is likely to show that the same people keep on being re-admitted. In one sense this is a failure of the processes of psychiatry, but in another sense each (carefully planned) discharge is a success, and a complex infrastructure often exists for maintaining the patient in the community. These include group support meetings, group therapy sessions, and social trips out of the hospital. We all have skills in the simple aspects of daily living, and in re-teaching these skills to our patients we may enable them to take the first steps in rebuilding their lives after a serious mental illness.

So *time*, *listening*, and the *skills of daily living* are our chief tools, and with these simple devices much can be done to rebuild the bridges between the patient and his outside world. These skills are simple compared with some of the highly elaborate skills such as psychoanalysis and hypnosis for which psychiatry is famous. The point of bringing them to the fore is so that the newcomer to psychiatry need not feel that there is a great weight of theoretical work to get through before he starts doing psychiatry. On the contrary, you can engage in the central process of psychiatry from your first day on the wards—using the knowledge and experience gained as a foundation on which to build the constructs required for the more specific and effective forms of psychotherapy.

In judging others, we expend energy to no purpose... But if we judge ourselves, our labour is always to our profit. Thomas à Kempis

If we knew more about ourselves, we would understand more about our fellow patients, bearing in mind that 'all gods and devils that have ever existed are within us as possibilities, desires, as solutions'. (Hermann Hesse, *Reflections*). What we want our patients to achieve is insight—and judging our fellows does not help this process. Judgment turns our patients away from us. We cannot expect our patients to be honest with us if they know that we are judging them.

- The good listener is *not* silent—but reflective. We hold a mirror up to our patients that they may get insight into themselves. Mirrors do not judge but they can enable self-judgment. Unless some criminal act is underway, it really does not matter what we think about our patients. What matters is how the patient thinks about him- or herself and his or her near-ones—and how these thoughts can be transformed.
- If we judge people they will not trust us. No trust \approx no healing.
- If we judge, patients will leave us for others less well-qualified.
- There is no evidence that judging improves outcomes. Worse outcomes are likely if the patient feels alienated.
- If patients detect that we feel bad about them, they may internalize this, and assume that things will always be bad because *they* are bad.

Notwithstanding all these bullets, there is a problem that will not go away. If we find ourselves talking to perpetrators rather than victims, we may not be wise to suspend judgment forever. If a crime is being committed putting others at risk, you may need to break confidentiality. Discuss this with a colleague. Ask yourself whether Nazi and Rwandan doctors were *too* nonjudgmental with their officer-patients. If '*tout comprendre c'est tout pardonner*', then to pardon *all* actions is to abdicate our moral selves. What is the consequence of this—for us, and for our patients? Unless we exercise judgment, it might be thought, we may be condoning evil. 'For evil to flourish in the world, all that is required is for the good to remain silent'. If we remain silent long enough, then will our own moral sense sicken, and die? What human duties do doctors have which trump *anything* that goes on in the consulting room? Whenever you think the time may have come to judge, check with yourself that it is not from outrage, or disgust, or through the exercise of pride, or from a position of power that you are judging—but reluctantly, and from *duty*. The dreadful history of some doctors in the twentieth century teaches that we must be human first, and physician-scientists second.

Listening

Contrasting dicta *Don't just do something—Listen!* (The Storr dictum)

When people are anxious it isn't clever to make a virtue of listening.

(The Parris dictum)

Sometimes our patients (and we ourselves) simply need to be **told**. Sometimes they need space for **self-expression**. Which approach is right—when? We only know this by knowing our patients: this entails listening, which is why, 9 times out of 10, Storr is right: but 9 out of 10 is not *always*.

Once we had the good fortune to work on an acute psychiatric ward with a would-be surgeon, who, before he accustomed himself to the ways of psychiatry would pace aimlessly up and down the ward after he had clerked in his patients, wondering when the main action would start, impatient to get his teeth into the business of curing people. What he was expecting, no doubt, was some sort of equivalent to an operating list, and not knowing where to find one he was at a loss, until he gradually realized that taking a history from a psychiatric patient is not a 'pre-op assessment', but the start of the operation itself. Even quite advanced textbooks of psychiatry appear to have missed the insight of this surgeon, describing psychotherapy as something which should only happen after 'a full psychiatric history' and after assessing what aspects of his problems are to be dealt with in treatment. There is no such thing as a full psychiatric history. In describing the salient psychological events of a single day even the best authors (eg James Joyce in *Ulysses*) require substantial volumes.

Taking a history sounds like an active process, and suggests a list of questions, and the format of our page on this process (p 322) would seem to perpetuate this error. It is not a question of taking anything. It is more a question of receiving the history, and of allowing it to unfold. If you only ask questions, you will get only answers as replies.

As the history unfolds, sit back and listen. Avoid interruptions for the first 3min. Be prepared for periods of silence. If prompts are needed try '... and then how did you feel?' or just '... and then. . .'; or repeat the last words the patient spoke. Do not be overanxious if the patient appears not to be covering major areas in the history. Lead on to these later, by taking more control as the interview unfolds. At first you will have to ask the relevant questions (p 322) in a rather bald way (if the information is not forthcoming during the initial unstructured minutes), but it is important to go through this stage as a prelude to gaining information by less intrusive methods. Always keep in mind the chief aims of *making a diagnosis*, defining problems, and establishing a therapeutic relationship.

What is the point of all this listening? Listening enables patients to start to *trust* us. Depressed patients often believe they will never get better. To believe that they *can* get better, patients need to trust us, and this trust is often the start of the therapeutic process. In general, the more we listen, the more we will be trusted. Our patients' trust in us can be one of our chief motivations, at best inspiring us to pursue their benefit with all vigour. A story bears this out. One summer's day, in 334 bc, Alexander the Great fell ill with fever. He saw his doctor, who gave him a medicine. Later he received a letter saying his doctor was poisoning him as part of a plot (it was an age of frequent, and frequently fatal intrigue). Alexander went to his doctor and drank the medicine—then gave him the letter. His confidence was rewarded by a speedy recovery. We think it is unreasonable to expect *quite* this much trust from our patients, and one wonders what can have led Alexander to such undying trust in his doctor. We strongly suspect that such a doctor, above all else, must have been a good listener.

Lifeworlds, and how to keep them intact

Even if we all listen the same way, what we will hear will depend on our own expectations, anxieties, and past experience. Take this dialogue:¹

Doctor: 'How long have you been drinking that heavily?'

Patient: 'Since I've been married'

Doctor: [impatiently] 'How long is that?'

Patient: [giggling] 'For years'. Perhaps the doctor hears '4 years'.

Perhaps the doctor is worrying about the length of time her liver has been exposed to excessive alcohol, or perhaps he just wants his notes to look complete. He needs facts in historical 'linear' time, but the patient chooses to answer in terms of event-time, or personal time. This is Mishler's great distinction between the 'voice of medicine' and the 'voice of the lifeworld'.² The doctor above has forced his patient to submit to his own world. Sometimes this may seem unavoidable: the chief thing is to recognize when this is happening, and to be preemptive by biting your tongue. What would this patient have gone on to say if her doctor had not time-raped her? What did those giggles signify? We will never know, but they might have explained her coming death.

¹ G Elwyn 1999 *BMJ* i 186 www.bmj.com/cgi/content/full/318/7177/186 ² E Mishler 1984 *The Discourse of Medicine: Dialectics of Medical Interviews*, Norwood, NJ, Ablex

Eliciting the history

Introduce yourself, and explain how long the interview will take. Describe its purpose. Find out how the patient came to be referred, and what his expectations are (eg about treatment). If the patient denies having any problems or is reluctant to start talking about himself, do not hurry him. Try asking 'How are you?' or 'What has been happening to you?' or 'What are the most important things?' Another approach for hospital patients is to indicate why the GP referred the patient and then ask what the patient thinks about this. Sit back and listen, without interrupting, noting exact examples of what he is saying. Take more control after ~3min, to cover the following topics.

Presenting symptoms Agree a problem list with the patient early on, and be sure it is comprehensive, eg by asking 'If we were able to deal with all these, would things then be all right?' or 'If I were able to help you, how would things be different?' Then take each problem in turn and find out about onset, duration, effects on life and family; events coinciding with onset; solutions tried; reasons why they failed. The next step is to enquire about mood and beliefs during the last weeks (this is different from the mental state examination, p 324, which refers to the mental state at the time of interview). Specifically check for:

Depression—the more specific these are, the greater the danger. Discussing suicide does not increase the danger. Questions to consider: 'Have you ever felt so low that you have considered harming yourself?' 'Have you ever actually harmed yourself?' 'What stopped you harming yourself any more than this?' 'Have you made any detailed suicide plans?' 'Have you bought tablets for that purpose?' **Thoughts**—ie low mood, anhedonia (inability to feel pleasure), self-denigration ('I am worthless'; 'Oh that I had not been born!'), guilt ('It's all my fault'), lack of interest in hobbies and friends plus biological markers of depression (early morning waking, decreased appetite and sexual activity, weight loss); **Delusions** (p 354); symptoms of **Psychosis** (persecutory beliefs, delusions, hallucinations, p 316); **Mania** and **Depression**; **Obsessive-compulsive disorder**; **Phobias**; **Personality disorder** (eg in young women; often not volunteered, and important). Note **Compulsions** (eg excessive hand-washing).

Present circumstances Housing, finance, work, marriage, friends.

Family history Physical and mental health, job, and personality of members. Who is closest to whom? Any stillbirths, abortions?

Birth, growth and development How has he spent his life? Ask about school, play (alone? with friends?) hobbies, further education, religion, job, sex, marriage. Has he always been shy and lonely, or does he make friends easily? Has he been in trouble with the law? What stress has he had and how has he coped with it? NB: noting early neurotic traits—nail-biting, thumb-sucking, food fads, stammering (not really a neurotic feature)—rarely helps.

Premorbid personality Before all this happened, how were you? Happy-go-lucky, tense, often depressed? Impulsive, selfish, shy, fussy, irritable, rigid, insecure? Talk to whomever accompanies him (eg wife), to shed light on premorbid personality and current problems. But don't let her speak for the patient (at least make sure that the patient has the first and last word).

Next examine the mental state (p 324 & p 353). You may now make a diagnosis. Ensure that the areas above have been covered in the light of this diagnosis so that the questions 'Why did he get into this way at this time?' and 'What is the best way to help him?' are answered.

Discussing childhood sexual abuse (CSA) with an adult who is currently experiencing psychological difficulties

This is a problem with many facets: a frequent question to arise is 'how far does past CSA account for current problems, and how much should this issue be explored now?' Each person is different: try to learn to use whatever your patient gives you, for their benefit. See p 328 for how to talk about sexual issues. Sometimes it is possible to be optimistic, despite the fact that many patients and professionals believe that CSA causes intense harm pervasively in the general population, regardless of gender. This issue has been examined in careful meta-analyses looking at 59 studies based on college data. These show that students with CSA were, on average, only slightly less well adjusted than controls. But this poorer adjustment cannot be attributed to CSA: family environment is consistently confounded with CSA and explains much more adjustment-variance than CSA. Self-reported reactions to and effects from CSA indicate that negative effects are neither pervasive nor often intense. Also, men react much less negatively than women. Even though this college data are consistent with data from national samples, this optimistic meta-analysis should not blind you to the possibility that the patient sitting in front of you might be very damaged by CSA—but do not assume that CSA is the underlying reason for everything.

medline:9670820

When in doubt, get further help.

The mental state examination

This assesses the state of mind at the time the interview is being conducted. Make records under the following headings.

- **General appearance & behaviour** Eg signs of self-neglect; excessive slowness; signs of anxiety or suspiciousness.
- **Manner of speech** Include the rate of speech, eg gabbling (pressure of speech), or slow and retarded. Note its content.
- **Mood** Note thoughts about harming self or others. Gauge your own responses to the patient. The laughter and grandiose ideas of manic patients are contagious, as to a lesser extent is the expression of thoughts from a depressed person.
- **Beliefs** Eg about himself, his own body, about other people and the future. Note abnormal beliefs (delusions) eg that thoughts are overheard, and ideas (eg persecutory, grandiose). See p 316.
- **Clauses, experiences or hallucinations** Note modality, eg visual.
- **Orientation** In time, place, and person. What is the date? What time of day is it? Where are you? What is your name?
- **Short-term memory** Give a name and address, and test recall after 5min. Make sure that he has got the address clear in his head before waiting for the 5min to elapse.
- **Long-term memory** Current affairs recall. Who is the Monarch? (This tests many other CNS functions, not just memory.)
- **Concentration** Months of the year backwards; see p 353.
- Note the patient's insight and the degree of your rapport.

Non-verbal behaviour Why are we annoyed when we blush, yet love it when our friends do so? Part of the answer to this question is that non-verbal communication is less well controlled than verbal behaviour. This is why its study can yield valuable insights into our patients' minds, particularly when analysis of their spoken words has been unrevealing. For example, if a patient who consistently denies being depressed sits hugging himself in an attitude of self-pity, remaining in a glum silence for long periods of the interview, and when he does speak, uses a monotonous slow whisper unadorned by the slightest flicker of a gesticulation or eye contact—we are likely to believe what we see and not what our patient would seem to be telling us.

Items of non-verbal behaviour

- | | |
|----------------------------------|----------------------------------------------|
| ● Gaze and mutual gaze | ● Dress: 'the apparel oft proclaims the man' |
| ● Facial expression | ● Hairstyle |
| ● Smiling, blushing | ● Make-up |
| ● Body attitude (eg 'defensive') | ● Body ornament |
| | (eg ear-rings, tattoos) |

Signs of auditory hallucinations: Anxious behaviour:

- | | |
|------------------------------------------------------------------------------------------------|--------------------------|
| ● Inexplicable laughter | ● Fidgeting, trembling |
| ● Silent and distracted while listening to 'voices' (but could be an 'absence' seizure, p 266) | ● Nail-biting |
| ● Random, meaningless gestures | ● Shuffling feet |
| | ● Squirming in the chair |
| | ● Sits on edge of chair |

Signs of a depressed mood:

- | | |
|---------------------------------|-----------------------------------------|
| ● Hunched, self-hugging posture | ● Downcast eyes; tears |
| ● Little eye contact | ● Slowness of thought; speech; movement |

What is a mental state?

A true description of a mental state entails valid knowledge about a person's current emotions plus his or her reactions to those emotions. These reactions are themselves emotional (eg being annoyed or dumbfounded at finding oneself in love again)—as well as being the bedrock out of which beliefs and attitudes are formed. These interactions make a picture to an external observer which is complex, paradoxical, subjective, error-prone, contradictory—and fascinating.

Describing and communicating mental states is the central puzzle that confronts not just psychiatrists and their patients, but also all artists. All the great poets and songwriters can summon up and make sense of diverse mental states (herein lies their genius)—but none can control them or their infinite progeny (what happens next). This might be said to be the proper province of psychiatry. If doctors could control mental states at will, then it is not necessarily the case that the job of psychiatry is done (there remains the problem of the will)—but at least the job would be half done (and, no doubt, there would be some unfortunate side-effects). Drugs, psychotherapy, and behavioural methods are the chief tools available for this task, and they all, crucially, impinge on mental state. You cannot tell if these modalities are helping if you cannot access your patient's mental state, which is why the page opposite is so important. But if you think you can access mental states *just* by applying the formulaic regimen opposite, you will often fail, because any trip into the mind of another is not just a voyage without maps, it is ultimately a creative and metaphysical enterprise.

On this view, knowledge of mental states is difficult and doubtful—but often this is not so: take for example the mental state of a baby being put to the breast after separation from his or her mother, or of an audience giving a standing ovation, or screaming fans waving to their idol, and we know without doubt that these mental states comprise unalloyed satisfaction, pleasure, and adulation. Note that so often it is non-verbal behaviour that allows valid judgment about mental states: don't rely on words alone—those capricious (but indispensable) tokens of disguise and deception.

Helping patients avoid doctor-dependency

A patient may become over-dependent on his or her doctor in many spheres of medicine. This is a particular danger in psychiatry because of the intimate and intense rather one-sided or asymmetrical relationship which may be built up between the patient and his psychiatrist—who is likely to come to know far more about the patient's innermost hopes and fears than many of his close friends. This encourages the patient to transfer to the therapist thoughts and attitudes that would more naturally be directed to a parent-figure. This process (known as transference) powerfully stimulates doctor-dependency, sometimes with serious consequences.

Signs of non-therapeutic dependency Repeated telephonings for advice, the inability to initiate any plan without help from the therapist, and the patient's disallowing of attempts by the therapist to terminate treatment (eg by threatening relapse).

Assessing whether dependency is a problem Clearly, in the examples above, the patient's dependency on his doctor is non-therapeutic. At other times, for example, early on in treatment, doctor-dependency may be quite helpful. In these circumstances the danger is that the doctor will be flattered by his patient's dependency on him. Most therapists either want to be loved by their patients or want to dominate them (or both), and it is important to know, in each session with each patient, just where you lie within the space marked out by these axes. Ask yourself 'Why do I look forward to seeing this patient?' 'Why do I dread seeing Mr X?' 'Why do I mind if this patient likes me?'

▶ When you feel good after seeing a patient always ask yourself why (it is so often because he is becoming dependent on you).

Avoiding dependency Planning and agreeing specific, limited goals with patients is one way of limiting dependency. If the patient agrees from the outset that it is not your job to provide him with a new job, wife, or family, the patient is more likely to have realistic expectations about therapy.

Planning discharge from the beginning of therapy is another important step in limiting doctor-dependency. Discharge is easy to arrange from the hospital outpatient department, but for the general practitioner the concept of discharge is diluted by the fact of his contractual obligation to the patient—who is perfectly within his rights to turn up the day after being 'discharged' and demand that therapy be started all over again. He must have more subtle methods at his disposal to encourage the patient to discharge himself. For example, he can learn to appear completely ineffective, so that the dependency cycle (patient presents problem → doctor presents solution → patient sabotages solution → doctor presents new solution) is never started. Another approach is to bore your patient by endlessly going over the same ground, so that the patient seizes control and walks out as if to say 'I've had enough of this!'

The foregoing makes patients out to be perpetual seekers after succour and emotional support—and so it can seem at times; but a great mystery of clinical medicine is that, spontaneously and miraculously, many apparently irremediably dependent patients *can* change, and start leading mature and independent lives. So do not be downcast when you are looking after these poor people: there is much to be said for simply offering a sympathetic ear, staying with your patient through thick and thin, and waiting for time to go by, and for the wind to change. Of course, the wind may change back again, but, if it does, you will not be back at square one, for you will be able to inject the proceedings with the most powerful psychotherapeutic agent of all: namely hope.

How to talk about sex with young people

Sexual issues may be an overt part of consultations relating to contraception, pregnancy concerns, and questions about sexually transmitted infections, and a covert part of those relating to emotional and behavioural difficulties, eg if there is abuse. So it is often relevant to take a sexual history. Doctors (and patients) can find this embarrassing, but this page may help to make consultations more effective (and less awkward).

The language used is very important. Sexual language can be categorized as follows: 1) Medical/technical (eg 'coitus'); 2) Slang (eg 'fucking') and 3) Socially acceptable (eg 'having sex'). It is not advisable to use slang as young people will think you are trying to be hip. Most will expect appropriate language; slang may shock them and may put up barriers. But occasional mirroring of the patient's words can be of help in gaining rapport. It is best to use socially acceptable terms, as these create a more informal and relaxed discussion than medical terms. Sometimes it is helpful to check out whether the young person finds your choice of word acceptable.

- Avoid ambiguity. Make sure that you both know what the other is talking about. For instance, some young people do not count oral sex as sex. So if asked 'have you had any sexual relations with her?' he may say no even though they had oral sex. If a new phrase crops up (slang changes all the time), ask for an explanation right away (a little gentle helping on your part usually overcomes any embarrassment here).
- Do not assume sexual knowledge. It is easy for adults to assume that media-exposed young people know everything, but this is not true. Just as when we were young, sex can seem confusing and mysterious. There are still many behind-the-bike-sheds myths, and it is just as hard as it ever was for young people to admit that they don't know something. Sex education in schools is uneven, and is sometimes useless.
- Don't appear embarrassed. It is easier for young people to open up if they think that you aren't going to blush, tell them off, or, worst of all, laugh. Don't act shocked and don't judge; give the wrong impression on this and they will stop being honest with you—see p 319 for discussion.
- Act as if you have plenty of time to listen. It is hard to get started when talking about personal feelings, and if the young person feels that their listener is impatient they may never begin at all.
- The more you practice sexual dialogue, the easier it becomes. If you shy away from it, it will remain a problem to you. Also, your patients may learn techniques of sexual dialogue, helpful in their lives as a whole, augmenting self-esteem, enabling sexual negotiation (useful in conflict resolution and negotiating safer sexual practices with partners). *medline 10938913* Also, you may be laying the foundation for honest sexual dialogue between this teenager and his or her offspring, 10–15yrs from now.

Asking about sexual abuse There are many different approaches:

- Have you been in any relationships that made you feel uncomfortable?
- Has anyone touched you in a way that made you feel embarrassed?
- I am wondering if anyone has hurt you in a sexual sort of way.

Explain the extent of the confidentiality that you are able to offer. The young person needs to be aware that you will have to discuss the contents of your conversation with someone else if you think that they or someone else is in danger. Get some printed information which can be used as a basis for discussion or can be given out at the end of a session.

Family Planning: 2 Pentonville Rd, London N1 9FP 0207 837 4044; Brook Centre: 165 Grays Inn Rd, London WC1X 8UD 0207 713 9000. Sex Education Forum: National Children's Bureau, 8 Wakley Street, London EC1V 7QE; Contraceptive Education Service 0207 837 4044

We thank Beth Miller & Justin Williams for formulating this page.

How to identify and use the full range of psychiatric services

Any good psychiatric service will have much more to it than out-patient clinics and hospital wards. The latter are very important, but many significant psychiatric interventions go on in the community. Alternatives to the cycle of outpatient/inpatient treatments include:

- Responsive community psychiatric teams
- On-call community psychiatric nurses (CPNs)
- Crisis intervention teams
- Substance abuse teams
- Drop-in services during weekends and unsocial hours
- Telephone helplines
- Self-help groups
- Mental illness hostels
- Occupation centres (voluntary or statutory)
- Sheltered work
- Supervised living quarters so patients are not alone
- Self-running group homes

These components of any comprehensive service are not only important in keeping patients out of hospital (a prime aim of those who are purchasing care—as well as of patients and doctors); they are also important in gradually normalizing the patient's relationship with his environment.

Integrated care has theoretical advantages—eg for a schizophrenic patient who is a substance abuser, integrating psychiatric and substance abuse therapy leads to better outcome than either alone. medline:11525078

In a way, the benefits of integrative care seem obvious (which is why it is puzzling that earlier meta-analyses have not found it to be advantageous, perhaps because the wrong questions were asked). medline:11034697

How to help patients not to be manipulative

Fireships on the lagoon We have all been manipulated by our patients, and it is wrong to encourage in ourselves such stiffness of character and inflexibility of mind that all attempts by our patients to manipulate us inevitably fail. Nevertheless, a patient's manipulative behaviour is often counter-productive, and reinforces maladaptive behaviour. A small minority of patients are very manipulative, and take a disproportionate toll on your resources, and those of their family, friends, and colleagues. We are all familiar with these patients who Ford Madox Ford describes as being like fireships on a crowded lagoon, causing conflagration in their wake.¹ After destroying their family and their home we watch these people cruise down the ward or into our surgeries with some trepidation. Can we stop them losing control, and causing melt-down of our own and our staff's equanimity? The first thing to appreciate is that, unlike an unmanned ship, these people *can* be communicated with, and you *can* help them without resorting to hosing them down with cold water.

Setting limits One way of avoiding becoming caught up in this web of maladaptive behaviour is to set limits, as soon as this behaviour starts. In a small minority of patients, the therapist may recognize that their needs for time, attention, sedation, and protection are, for all practical purposes, insatiable. Whatever the therapist gives, such patients come back for more and more, and yet in spite of all this 'input' they do not appear to get any better. The next step is to realize that if inappropriate demands are not met, the patient will not become sicker (although there may be vociferous complaints). This realization paves the way for setting limits on the patients' behaviour, specifying exactly what is and is not acceptable.

Take for example the patient who demands sedation, threatening to 'lose control' if it is not given immediately, stating that he cannot bear living another day without sedation, and that the therapist will be responsible for any damage which ensues. If it is decided that drugs do not have a part to play in treatment, and that the long-term aim is for the patient to learn to be responsible for himself, then it can be simply stated to the patient that medication will not be given, and that he is free to engage in destructive acts, and that if he does so this is his responsibility.

The therapist explains that in demanding instant sedation he usurps her professional rôle, which is to decide these matters according to her own expert judgment, and that such usurpation will not be tolerated. If there is serious risk of real harm, admission to hospital may be indicated, where further limits may be set. If necessary, he is told that if he insists on 'going crazy' he will be put in a seclusion room, to protect others.

¹ Ford Madox Ford 1915 *The Good Soldier*, Penguin

See also J Holmes & A Elder 2002 *Mental Health in Primary Care*, oup

How to help patients be less lonely

Being unwanted is the worst situation that any human being can experience. Loneliness does not come from having no people about one, but from being unable to communicate the things that seem important to one.

Loneliness is not a condition you will find in many psychiatric classifications—yet it is the root of much psychopathology. It is also frequently the result of psychopathology. Some people, of course, like to avoid their fellows. For them, intrusion into their private world may cause despair; but, for most of us, these intrusions are welcome, and necessary for health. Loneliness seriously affects 1 in 10 older people, and can contribute to alcoholism, depression, and suicide. Suspect that loneliness is important whenever you find your hand being gripped for comfort after one would have thought that a social encounter was over—also whenever there is a verbal outpouring and a 'defeated demeanour'.¹

Do not assume that loneliness means social isolation. Someone who has brief visits 3 times a day, from, say, a nurse, a care assistant, and a 'meals on wheels lady' is not socially isolated, but may be very lonely. But if he or she gets on well with just one of these visitors, this can be enough to banish loneliness. So this is the first lesson: *be nice to people, and take trouble to find out their hopes and fears*. But more is possible: in general, it is depressing waiting for the doorbell to ring, so tailor your suggestions in the light of your patient's mobility. This needs initiative on your patient's part: but do not think you must treat your patient's depression *before* you tackle a lonely lifestyle: tackling this may be the route out of depression. Areas to think about include:

- What facilities already exist? Is there a local pub, day centre, or lunch club available? 'God, I wouldn't be seen dead in one of those places!' we so often hear. But take time to point out that it does not matter initially whether they get anything out of a social interaction. After all, they may meet someone of like mind, so enabling these artificial crutches to be thrown away.
- Is the person religious? There may be activities and outings to plan and talk about, and reminisce over, even if not actually enjoyed at the time.
- Housing: if the person is planning a move, will they be near family, and other people who speak their own language (metaphorically and literally)?
- Alternative therapies, eg massage and aromatherapy, can relieve loneliness.
- Adult education classes are a good (but often expensive) way to meet like-minded people—and with the acquisition of new skills, confidence improves and social interaction becomes more pleasurable.
- Involvement with community action groups may be a source of friends (and a source of frustration and disappointment—but do not expect your interventions to be without side-effects: the thing is to plan for them).
- Details of local community activities can be found in the UK at the local *Council for Voluntary Services*. Other organizations advertise at local libraries.
- While at the library, ask about joining a book club.
- Befriending schemes can be very helpful to those who are housebound.
- Technology forums such as the Internet may provide relief from boredom and loneliness—and for some this will offer the best chance of meeting with a kindred spirit, unlimited by the constraints of time and space.
- Befriending others, and offering phone support is an option, whether or not one is housebound. Some local authorities give help to get phones.

• Mother Teresa : Carl Jung 1 A Forbes 1996 *BMJ* ii 352. **Useful UK addresses:**

University of the Third Age, 26 Harrison St, London WC1 tel. 020 7837 8838

Names of leaders of local religious groups: 15 Henrietta St, London WC2E 8QH, tel. 020 7240 0630

Age Concern: 1268 London Road, London SW16 4EJ, tel. 020 8679 8000

Cruse: Bereavement line: 020 8939 9530

Gaga Holidays: Bouverie House, Middelburg Square, Folkstone, Kent CT20 1AZ

How to recognize and treat your own mental illnesses

Doctors have a higher than average incidence of suicide and alcoholism, and we must all be prepared to face (and try to prevent) these and other health risks of our professional and private lives. Our skill at looking after ourselves has never been as good as our skill at looking after others, but when the healer himself is wounded, is it clear that his ability to help others will be correspondingly reduced? Our own illnesses are invaluable in allowing us to understand our patients, what makes people go to the doctor (or avoid going to the doctor), and the barriers we may erect to resist his advice. But the idea of an ailing physician remains a paradox to the average mind, so that we may ask: ►Can true spiritual mastery over a power ever be won by someone who is counted among her slaves?¹ If the time comes when our mental state seriously reduces our ability to work, we must be able to recognize this and take appropriate action. The following may indicate that this point is approaching:

- Drinking alcohol before ward rounds or surgeries.
- The minimizing of every contact with patients, so that the doctor does the bare minimum which will suffice.
- Inability to concentrate on the matter in hand. Your thoughts are entirely taken up with the workload ahead.
- Irritability (defined as disagreeing with >1 nurse/24h).
- Inability to take time off without feeling guilty.
- Feelings of excessive shame or anger when reviewing past débâcles. To avoid mistakes it would be necessary for us all to give up medicine.
- Emotional exhaustion (defined as knowing that you should be feeling pleased or cross with yourself or others, but on summoning up the affairs of your heart, you draw a blank).
- Prospective studies suggest that introversion, masochism, and isolation are important risk factors for doctors' impairment.

The first step in countering these unfavourable states of mind is to recognize that one is present. The next step is to confide in someone you trust. Give your mind time to rejuvenate itself.

If these steps fail, various psychotherapeutic interventions may be relevant, eg cognitive behavioural therapy (p 370), or you might try prescribing the symptom. For example, if you are plagued by recurring thoughts about how inadequately you treated a patient, set time aside to deliberately ruminate on the affair, avoiding distractions. This is the first step in gaining control. You initiate the thought, rather than the thought initiating itself. The next step is to interpose some neutral topic, once the 'bad' series of thoughts is under way. After repeated practice, the mind will automatically flow into the neutral channel, once the bad thoughts begin, and the cycle of shame and rumination will be broken.

If no progress is made, the time has come to consult an expert, such as your general practitioner. Our own confidential self-help group for addiction and other problems is the British Doctors' and Dentists' Group and may be contacted via the Medical Council on Alcohol (uk tel. 020 7487 4445). If you are the expert that another doctor has approached, do not be deceived by this honour into thinking that you must treat your new patient in any special way. Special treatment leads to special mistakes, and it is far better for doctor-patients to tread well-worn paths of referral, investigation, and treatment than to try illusory short cuts.

Burnout

Risk factors—for doctors Pressure of work, a less personal relationship with patients, overly formal hierarchies, and suboptimal income are put forward to explain the fact that some doctors (urologists, for example) in the public sector are more at risk of burnout compared with private-service urologists. medline:11257659

Signs Ask about: motivation, exhaustion, and depersonalization; find out about intentions to relocate (where, exactly? in the life hereafter?). Also:

- Stress •Depression •Libido↓ •Insomnia •Guilt •Feeling isolated
- Apathy •Paranoia •Amnesia •Indecision •Denial •Temper tantrums

Management (Difficult) Some may respond to plans such as these:

- Diagnose and treat any depression (p 336–42).
 - Allow time for the person to recognize that there is a problem.
 - More hobbies, and more nice holidays.
 - Advice from wise colleagues in the specialty (regular follow-up).
- Mentoring consists in forming a supportive relationship with an independent colleague for the sole purpose of support.
- Learn new professional skills.
 - Set achievable goals in work & leisure (eg protected time with family).
 - Early retirement.

Prevention Strategies such as career counselling are said to be effective but really do no more than point a lollipop at a furnace. medline:11311835

Patients in disgrace: the fat-folder syndrome

One of the rôles of the psychiatrist is to act as the terminus for patients who have been shunted from hospital department to hospital department. These patients offer their doctor an 'organ recital' of incapacitating physical symptoms for which no physical cause can be found (somatization). She knows that 'there must be something wrong'. In the end, the patient is sent in disgrace to the psychiatrist because the doctor declares that there is 'nothing wrong' and that the patient is 'just a hypochondriac'. The psychiatrist then has to do much work undoing the cycle of symptom-offering and investigation, by acknowledging that there is something wrong. The aim is to reframe symptom-offering into problems which need to be solved, starting with the doctor accepting that the patient is distressed and looking for helpful responses from her carers which are yet to be defined. After establishing rapport, the doctor agrees a contract with the patient, that he will give regular consultations for listening to how the patient feels and will try to offer help, if she acknowledges that past investigations have not helped, and that she agrees not to consult other doctors until a specified number of sessions have elapsed. In addition, cognitive therapy (p 370) which examines the way that conscious thoughts and beliefs perpetuate disability, can lead to meaningful symptom reduction.

medline: 11280341

Dissociation (formerly known as hysteria)

Our stream of consciousness does not progress from cradle to grave as a single line: there are separations (dissociations) and confluences—for example, when we day-dream, or drive to Porlock with no recollection of the scenery along the way—only of our inner landscape. Dissociation may be partially adaptive, eg in annihilating pain in near-death experiences. Another example is a patient who was homosexually raped, had no conscious memory of the event, but felt an irresistible urge to write insulting letters about the person who had raped him. Most instances subside spontaneously, but if they do not it is important to refer early to a psychiatrist, before associated behaviour becomes habitual.

Types of dissociation *Amnesia* is the commonest type: see OPPOSITE. *Depersonalization* (Feeling of being detached from one's body or ideas, as if one were an outsider, observing the self; 'I am in a dream' or 'I am an automaton'—but unrelated to drugs/alcohol). This may be a response to stress.

Dissociative identity disorder The patient has multiple personalities which interact in complex ways. It is present in 3% of acute psychiatric inpatients. *Identity disturbance* Inability to recall one's past ± loss of identity or formation of a new identity—associated with unexpected, purposeful travel (lasts hours to months, and for which there is no recollection).

Prognosis Follow-up over ~6yrs shows that ~5% of those referred to a CNS hospital who had hysteria or dissociation diagnosed turned out to have organic illness. However, these patients may not be typical of those diagnosed with dissociation (diagnoses included spinocerebellar degeneration).

Treatment Exploring the stresses in the patient's life may help. In general, be ready to recognize the psychological components of physical illness, and get expert psychiatric help, while leaving the door open for reappraisal if new features develop.

Is this amnesia dissociative?

- Has a physical cause been carefully discounted? (Drugs, epilepsy, etc.)
- Is the patient young? Beware making the first diagnosis if >40yrs old.
- Have the symptoms been provoked by stress? Ask the family.
- What is the pattern of amnesia? If for distant *and* near memories, then dissociation is more likely (as opposed to organic causes) than if the amnesia is for shorter term memory.
- Is there secondary gain?—ie is it worthwhile perpetuating symptoms? Many diseases have gain for the patient: this sign is most useful if *absent*.
- Do related symptoms 'make sense' (eg aphonia in a news-reader)?
- Indifference to what should constitute a major handicap (*la belle indifférence*) is of marginal diagnostic use.
- Is malingering likely? The answer is almost always 'No', except in prisons and the armed services (when secondary gain is easy to identify).
- Is there a dissociative personality? The dissociative experiences scale (DES) screens for this. It is a 28-item visual analogue scale on which patients indicate percentages of time during which they have different types of dissociative experiences (excluding those influenced by alcohol or drugs)—ranging from non-pathological experience of being so absorbed in tv that the person is unaware of what is happening in the room, to severe dissociations, eg of having no memory of important personal events, or feeling that his or her body belongs to another. Those with dissociative disorders typically have DES scores of 30 or higher; most others score nearer zero.¹

1 B Greyson 2000 *Lancet* 355 460

Calibrating our sensitivity to psychosomatic events

Why do some doctors preferentially diagnose somatic illness? Why, when confronted by unexplained symptoms do we often subconsciously try to fit them to a physical ailment? The reason is usually that prescribing a pill is easier than changing, or regulating, intrapsychic events. The patient and the doctor may collude with this approach, and then get very frustrated when it yields nothing of value. Alternatively, some doctors are so used to diagnosing psychological problems that they are all too prone to launch into treating someone's depression and malaise, rather than their endocarditis or brucellosis. There is no single correct approach. We all make errors—but the point is to find out in which direction you tend to make errors, then allow for this in your interactions with patients.

Depression

Each year 40% of us have quite severe feelings of depression, unhappiness, and disappointment. Of these, 20% experience a clinical depression, in which low mood occurs with sleep difficulty, change in appetite, hopelessness, pessimism, or thoughts of suicide. **Criteria** of major depression:

- Loss of interest or pleasure—**anhedonia** in daily life with dysphoric mood (ie 'down in the dumps') plus ≥ 4 of the following (the first 5 are 'biological' symptoms)—present nearly every day for at least 2 weeks:
- **Reduced energy** with **fatigue** (or, rarely, increased appetite).
- **Weight change**—with **weight loss** (worse in mornings).
- **Reduced or increased movement** (ie a paucity of spontaneous movement, or sluggish thought processes), or **psychomotor retardation**.
- Decrease in **sexual drive** and other appetites.
- Evidence of (or complaints of) reduced **concentration**.
- Ideas of **self-harm**, inappropriate **guilt** or **self-blame**.
- Recurrent **thoughts of death** and **suicide**, or **suicide attempts**.

Why is depression so often missed? • Lack of knowledge. • Preoccupation with physical disease. • Psychiatric labels are hated (doctors and patients collude). • It's hard to spot depression coexisting with other illness.

Classification Classify as: • **Mild; moderate; severe**. • **With/without biological features**. • **With/without delusions or hallucinations**. • **With/without manic episodes** (ie bipolar not unipolar). These replace the old reactive/endogenous labels. **Excludes** Cyclothymic disorder; substance-induced mood disorder; schizophrenia, dementia; mood disorder due to a general medical condition.

Why people get depressed: some ideas • **Genetics**: identical twins reared apart show 60% more concordance for depression than dizygotic twins, but this sort of twin study is very suspect; see [medline:11780600](#) for the reasons why.

- **Biochemistry**: there are excess 5-hydroxytryptamine (5HT₂) receptors in the frontal cortex of brains taken from suicide victims. See [OHCM](#) p 324.
- **Endocrinology**: cortisol suppression (dexamethasone suppression test, [OHCM](#) p 301) is abnormal in ~30%. Melatonin is also implicated (p 402). Also, 17 β -estradiol is known to help depression if perimenopausal. [EBM](#) 2002 77 21
- **Stressful events** (eg new births, job loss, divorce, illness): present in 60%.
- **Freudian reasons**: depression mirrors bereavement, but loss is of a valued 'object', not a person. There is ambivalence with hostility turned inwards.
- **Learned**: if punishment is unrelated to actions, but is perceived as random, the response is helplessness and depression. [medline-11000446](#)
- **Vulnerability factors**: physical illness, pain, and lack of intimate relationships may allow depression to arise and be perpetuated. [medline-11007705](#)

Management There is no clear distinction between the low moods we all get and illness needing vigorous treatment, but the lower the mood and the more marked the slowness, the more vigorous the treatment needs to be.

- **Cognitive therapy** (eg cognitive therapy, p 370) is part of the treatment of *all* depression; it may be all that is needed in milder depressions.
- **Presence of biological features or stress predicts a good response to drugs** (p 340) especially if symptoms are severe. Not everyone wants drugs. Discuss all options. Herbalism can work: hypericum (St John's wort) may be useful in mild to moderately severe depression. [medline-10796719](#)
- **Delusions or hallucinations prompt a physical treatment**, ie **ECT** (p 342) or **antidepressants** (antidepressants \pm antipsychotic drugs, p 360).
- **The depressive phase of bipolar illness is treated as above**, but note:
 - Be cautious with the physical treatments: they may induce mania.
 - Lithium or valproate prophylaxis may be needed (p 354). [medline 11313093](#)
- **Social circumstances**; high suicide drive; isolation.

Who is likely to benefit from antidepressant drugs? (p 340)

Those who have had low mood or loss of desire for pleasure (anhedonia) most of the day for at least the last 2 weeks who show 4 or more of the following 7 markers of severe depression:

- Suicide plans in existence, or ideas of self-harm.
- Unexplained or unbidden feelings of guilt or worthlessness.
- Inability to function—eg psychomotor retardation or agitation.
- Concentration impaired.
- Impaired appetite.
- Decreased sleep (especially early morning waking).
- Energy low, or unaccountable fatigue.

► Enquire about these phenomena *whenever* depression is possible.
NB: treatment may still be needed if the above criteria are not met: listen to their story. Patients will often not accept that they could be depressed because “there is nothing to *make* me depressed”. It helps to suggest that they could still be depressed, and that treatment could be very helpful. Give them time to go away and think about it, perhaps discussing it with someone they know. Try ‘would your wife (or partner) say you were depressed? Please could you ask and let me know what they say?’ Such patients often return enriched by dialogue and reflection, and are successfully treated. This may not be over-medicalizing a patient’s symptoms: there is evidence that such patients are simply inexperienced in understanding their depression: when they *next* become depressed, their views much more nearly match those of their doctor.

www.txt.co.uk/full.cfm?1093

Suicide and attempted suicide

Suicide needs to be distinguished from attempted suicide and self-harm. The latter is very common, but remember that every non-fatal event may be fatal next time, which is why the subjects are treated together here. Men aged 15–30 are most at risk of completed suicide; incidence of self-harm is highest in teenage and older females (>1%/yr). Suicide rates are rising (now as common as road deaths medline:11897274) being higher in island peoples and Europe, and rarer in Islamic peoples. Worldwide, 10⁶ people/yr commit suicide; increasingly they are young. wasserman 200
isbn: 1 85317 822 5; medline:11288165 & 10625249

Understanding suicide Suicide may be a way of avoiding pain, or keeping honour and autonomy, and avoiding shame (the noble Roman in us prefers suicide to being forced to walk on our knees). *Suicide may be a way of:*

- Controlling change in families, eg when a member is about to leave.
- Communicating important messages in a not-to-be ignored way.
- Gaining power in family transactions, by escalating conflict.

Often an argument with a partner is an antecedent to self-harm. Immaturity, inability to cope with stress, weakening of religious ties, and availability of drugs (psychotropics & alcohol are popular poisons) are also important, as is 'copy-cat' behaviour: when celebrities try suicide, others follow. Antecedents of suicide: chronic disease, depression, bankruptcy, etc. medline:7701505
Reducing levels of psychiatric care are also important. medline:10227220

Bullying, intolerable stress to succeed, and 'falling behind in my homework' are also risk factors. If this mirrors your own state after trudging through too many pages, shut this book, and have a good holiday.

After assessment, there are 5 stages in trying to help the survivors:

- Agreement of a contract offering help (p 339)—by negotiation.
- Discussion with the family as to how problems might be tackled.
- Problem-solving by facilitating the patient's understanding of her predicament, and by pointing out how she has coped with problems in the past. The aim is to engender a greater ability to cope in the future as well as helping immediate personal and social problems.
- Prevention: open-access, walk-in clinics or 24h phone service.
- Follow-up—either with family or with the patient alone.

Assessment Think of a target with 3 concentric rings. The *inner ring* is the circumstances of the attempt: what happened that day; were things normal to start with? When did the feelings and events leading up to the act start? Get descriptions of these in detail. Was there any last act (eg a suicide note)? What happened after the event? Was this what he/she expected?

The *middle ring* is the background to the attempt: how things have been over the preceding months. Might the attempt have been made at any time over the last months? What relationships were important over this time?

The *outer ring* is the family and personal history (p 322).

After the above, come to the *core*—the intentions lying behind the attempt, and the present feelings and intentions. Does the attempt represent a wish to die (a grave, not-to-be-ignored sign); a wish to send a message to somebody; or a wish to change intolerable circumstances? Ask: 'If you were to leave hospital today, how would you cope?' Examine the mental state (p 324) to find out if there is any mental illness. **Summary** •Any plan? What? When? Where? •Are the means available? •Ever tried before? How seriously? •Preparations (making a will, giving things away).

Before arranging hospital admission, ask what this is for. Is it only to make you feel happier?—or to gain something that cannot be gained outside hospital. Ask: *Why will discharge be safer in a few weeks rather than now?*

How to cope with threats of suicide

Suicidal behaviour is powerful. *Boyfriend*: 'After last night, I think we should separate—we'd better just be good friends. . . ' *Girlfriend* thinks: 'You are not going to treat me like that', but says: 'So we are not getting married after all?' 'Well not for a little while, maybe.' 'So you don't love me . . . I knew you loved Amanda more than me.' Exit to bathroom, where she swallows handfuls of her mother's tranquillizers. Next day in hospital the boyfriend says 'I'm so sorry—I didn't know you loved me that much.' He thinks: 'No one has ever thought that much of me. . . Fancy wanting to kill herself because I wouldn't marry her! I feel guilty; I should not have led her to this. . . ' Whereupon he proposes marriage. This disastrous ending may be complicated by threats of suicide from jilted Amanda.

The psychiatrist may become enmeshed in these webs of suicide threats, and may wrongly assume that because someone threatens suicide, they should be admitted to hospital (compulsorily if necessary) so that they can be kept under constant surveillance, and suicide prevented. This reasoning has 3 faults. The first is the idea that it is possible to prevent suicide by admission. There is no such thing as constant surveillance. Second, hospital admission may achieve nothing if it simply removes the patient from the circumstances which he must learn to cope with. Third, it is necessary to distinguish between suicide gestures, which have the object of influencing others' behaviour, and a genuine wish to die.

Before death, many suicide victims see a GP, and it is wise to be alert to undercurrents of suicide which only sometimes surface during consultations. Ask *unambiguously* about suicide plans (p 322). On deciding that a threat is more manipulative than genuine, very experienced therapists may influence the person's use of suicide behaviour by forcing him to face the reality of his suicide talk, eg by asking: 'When will you kill yourself?' 'How will you do it?' 'Who will discover the corpse?' 'What sort of funeral do you want? Cremation, burial, with or without flowers?' 'Who will come?'

The standard pointers to risk of death from a suicide attempt are being male, older, unemployed, with prolonged psychiatric or painful illness, ± drug abuse. Recent attempts to validate this list have failed. It is better to think dynamically of risks and protective factors (such as family support), with suicide occurring after key events which accumulate risk.

► *All suicide threats must be taken seriously*—but the emphasis will differ depending into which group the patient falls. In both groups it may be helpful to form a *contract* with the patient, eg:

- The therapist will listen and help if the patient agrees to be frank, and to tell the therapist of any suicide thoughts or plans.
- Agreement about which problems are to be tackled is made explicit.
- The type of change to aim for is agreed.
- Specification of who else will be involved in treatment (eg other members of the family, friends, the patient's GP).
- An agreement about the timing and place of sessions.
- An agreement about the patient's responsibility to work effectively with the therapist, and to carry out any 'homework'.

If in doubt about the energy with which we may pursue our own destruction, let us call to mind a notable Japanese pilot who, during World War II, persistently volunteered to be a Kamikazi pilot to run a suicide mission against us ships, in the defence of Okinawa. The authorities just as persistently refused his request—he had a wife and 3 daughters. He kept on reapplying, determinedly. Not wanting to risk her husband's failure again, and not wanting to stand in Destiny's way, his wife killed their 3 daughters, and then herself, so removing the obstacle to her husband's mission—and on May 28, 1945, he finally took to the air, and achieved his end.

Which antidepressant? Some sample regimens

Drugs improve mood and ↑ synaptic availability of norepinephrine or 5HT.

► Never rely on drugs alone. Your own personal qualities ± psychotherapeutic interventions are just as important. See p 368 for more details. Specifically encourage socializing, and the countering of negative thoughts.

● **Unrelieved major depression, no drug agent, risk suicide** If a cheap agent is essential, try dothiepin (a tricyclic) 50mg/8–24h po. Start with 50–75mg at night. Explain side-effects: sedation, dry mouth, urine retention, blurred vision, postural hypotension, ↑ glaucoma risk. Warn about driving and using machinery. Explain that benefits take weeks to develop. Avoid if risk of arrhythmia (eg post myocardial infarction). We have never been persuaded that any other drug is *more* effective than dothiepin. If side-effects are tolerated, it is an excellent drug. Typical dose: ≤150mg at night. Maximum: 225mg/day (eg in hospital use). medline:11059441

● **Depression with antidepressant** Try SSRIs, eg fluoxetine, 20mg/24h po. $t_{1/2} \approx 3$ days. In women, consider combining with 0.5mg/day folic acid: folate is low in major depression, and supplementation did improve response, in one randomized study. N=127
medline:10967371

● **Past history of good response to tricyclics, now suicidal** Try lofepramine, 70mg/12h (less likely to be fatal in overdose; less risk of fatal arrhythmias).

● **Depression with antidepressant** Tricyclics rarely help? try fluoxetine, above.

● **Depression with antidepressant** May be less tolerant of SEs: halve doses (tricyclics).

● **Depression with antidepressant** (p 342) ± neuroleptics (p 360) may be needed.

● **Depression with antidepressant, persisting on drug** Paroxetine (20mg each morning, increased by 10mg increments; max: 50mg/24h) is safer than tricyclics. It is the SSRI most associated with the unpleasant dystonias on withdrawal.

● **Depression with antidepressant, sleep pattern** Tricyclic, eg dothiepin, as above. If suicidal, nefazodone (SSRI, & blocks 5HT₂ receptors) and mirtazapine (blocks 5HT₂, H₁, and 5HT₃—15mg at bedtime, max 45mg) *may* have a rôle. Warn patients not to rely on the 'fact' that day time sleepiness usually wears off after a few weeks. NB: ordinary SSRIs can aggravate insomnia. Sleep is such a restorative for some patients, so give them the best chance.

● **Depression with antidepressant, eg SSRI** Venlafaxine *may* have a rôle. medline:11983635

● **Depression with antidepressant** Tricyclics may be best; p 408; get expert help.

● **Depression with antidepressant, eg SSRI** Clomipramine or an SSRI.

● **Depression with Parkinson's disease** Nortriptyline has been found to be effective, and may not make the Parkinson's much worse.

● **Depression with antidepressant** Nortriptyline is ?better than fluoxetine. medline:10698809

● **Depression with antidepressant, eg SSRI** HRT (17β estradiol) is helpful, p 18. EBM 2002 77 21

● **Depression with antidepressant** Fluoxetine sometimes leads to weight ↓. medline:9693356

● **Depression with antidepressant, eg SSRI** Offer psychotherapy. Does he think that drugs equate with moral failure? If still unconvinced, perhaps an alternative therapy such as St John's wort might be acceptable? meta-anal
medline-10796719

Duration ► Don't stop too soon. Go on for a few months after you achieve full remission. If you don't, expect a relapse. If there have been 3 or more previous depressions, consider continuous treatment. Always tail off doses slowly. Prepare the patient for life's ups and downs, and explain that these are not relapses, going back to square one. Arrange follow-up.

Receptors No evidence exists that antidepressants acting at >1 receptor sites differ in efficacy from SSRIs in major depression. meta-anal
www.txt.co.uk/full.cfm?1094

Electroconvulsive therapy (ECT)

Indications Depression if: •Not responding to antidepressants, or •Psychotic features are present, or •Needing to be controlled fast (eg patient refusing to drink, high suicide risk, or postpartum): electroconvulsive therapy (ECT) is the fastest acting physical treatment. Emergency ECT is possible, eg in some elderly patients, but should rarely be necessary.

Serious effective depression (depression with signs of schizophrenia).

Management If not responding to drugs. *Typical course length* 6 sessions (2/week)

Contraindications Recent subdural/subarachnoid bleeding; lack of consent (p 400; involve relatives, but they cannot consent for another adult).

Cautions Recent stroke, myocardial infarct, arrhythmias. SE: anterograde amnesia; anaesthesia complications.

Technique Check the patient's identity and that 'nil by mouth' for >8h.

- Ensure a detailed medical history and physical examination has been done, and any illnesses investigated and treated as far as possible. High anaesthetic risk?—See p 762; seizure threshold ↑ if on concurrent benzodiazepines, anticonvulsants, or methohexital (=methohexitone) >1.2mg/kg; also ↑risk of heart complications if on tricyclics. Liaise between psychiatric and anaesthetic staff. Do benefits outweigh risks?
- Give calm reassurance away from the site where ECT is going on. (It may be distressing to watch ECT actually happening.)
- Are the consent forms in order (p 400)? In the rare instances in the UK where ECT is given without consent, a second opinion from the Mental Health Commission must state that the treatment is necessary (p 400).
- Ensure that fully equipped resuscitation trolleys are present including a functioning defibrillator, suction apparatus, and pulse oximeter.
- Ensure that the anaesthetist (senior and ECT trained) knows about allergies or drugs interfering with anaesthesia. For countering ECT-induced vagal stimulation, she is most likely to use atropine or methscopolamine before using an ultra-short-acting anaesthetic agent (eg methohexital) with muscle relaxation (eg with suxamethonium) to minimize the seizure.
- The ECT machine: checked recently? Reserve machine to hand? What charge/energy is to be given? Which waveform will be used (bidirectional or modified sinusoidal, or unidirectional)? Study manufacturer's information.
- In unilateral ECT*, position one electrode (A) 4cm above the midpoint between the auditory meatus and the lateral angle of the eye, and the other (B) 10cm away from A, above the ear—see diagram opposite.
- Put electrode jelly on the chosen sites (not enough to allow shorting).
- When the anaesthetist gives the word, give the shock. Be prepared to restrain the patient if paralysis is incomplete. While the current passes, the muscles will contract. This will cease as the current ceases. After ~10sec, further clonic spasms occur, lasting about 1min. The only sign may be lid fluttering. Clonus is probably needed for ECT to be fully effective.
- Put the patient in the coma position. Check BP and pulse, etc until conscious. Ask the anaesthetist to consider iv midazolam for those (few) who become very agitated during the recovery process.

ECT may be frightening, partly from its bad press; so explain, step-by-step, what will happen; and patients should not witness others having ECT. The author also finds it helpful to tell patients that he would opt for ECT, if the right indications pertained, because it can be so effective—indeed life-saving.

*Right unilateral high-dose ECT is rarely used but is as effective as bilateral ECT, but produces less severe and persistent cognitive effects. medline:10807482

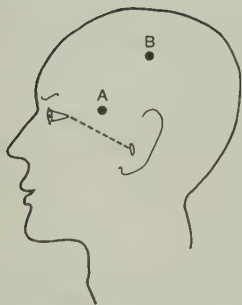
What is the correct 'dose' of unilateral ECT?

There is no universal answer, but note that there is some evidence that the therapeutic efficacy of ECT is proportional to the length of seizure induced. Be sure that you have adequate training on this issue by the consultant in charge of the session. Dose is better measured in millicoulombs (mC) than milliamps. It depends on *seizure threshold*, which varies 40-fold among patients. A moderately 'suprathreshold' dose (eg 200% above seizure threshold) usually gives seizures of adequate duration, while aiming to minimize cognitive side-effects—according to the Royal College of Psychiatrists *ECT Handbook*.

Doses need changing depending on response, and dose of methohexital (or propofol —[medline:11294464](#)) in the anaesthetic: also, seizure threshold rises by ~80% as courses progress. NB: a 'good' seizure is one which is of adequate duration (~15sec during early sessions; duration is less important later), with both a tonic and a clonic phase. Some machines allow EEG monitoring—useful as anaesthesia renders seizure analysis difficult. If the seizure lasts >2min, tell the anaesthetist, who will provide a bolus of diazepam, or more of the general anaesthetic agent.

The frequency and speed of response of ECT varies according to indication, eg bipolar depression patients show more rapid improvement and need fewer treatments than unipolar patients.

[medline:11333069](#)



Electrode positions (see text)

Anxiety neurosis/generalized anxiety disorder

Anxiety is a universal experience; it is, according to various reckonings, the chief factor limiting human potential; it causes much suffering, costing the UK \geq £5 billion/yr. *Neurosis* refers to **maladaptive psychological symptoms not due to organic causes or psychosis, and usually precipitated by stress**. Apart from free-floating anxiety and depression, such symptoms are: fatigue (27%), insomnia (25%), irritability (22%), worry (20%), obsessions, compulsions, and somatization (p 744). These are judged to be more intense than normal, ie out of proportion to the stress that precipitates them. Symptoms are not just part of a patient's normal personality, but they may be an exaggeration of personality: a generally anxious person may become even more so—ie develop an anxiety neurosis—as a result of job loss. The type of neurosis is defined by the chief symptom (eg anxiety, obsessional, depressive). Before diagnosing neurosis, consider carefully if there is underlying depression that needs treating with antidepressants. medline:11463676

Symptoms of anxiety Tension, agitation; feelings of impending doom, trembling; a sense of collapse; insomnia; poor concentration; 'goose flesh'; 'butterflies in the stomach'; hyperventilation (so tinnitus, tetany, tingling, chest pains); headaches; sweating; palpitations; poor appetite; nausea; 'lump in the throat' unrelated to swallowing (globus hystericus); difficulty in getting to sleep; excessive concern about self and bodily functions; repetitive thoughts and activities (p 346). **Children's symptoms:** Thumb-sucking; nail-biting; bed-wetting; food-fads. The various types of anxiety:

- Generalized anxiety disorder
- Panic disorder
- Simple phobia
- Post-traumatic stress disorder
- Agoraphobia
- Social phobia
- Obsessive-compulsive disorder.

Causes Stress (eg work—or lack of it, noise, a hostile home), and life events (eg gaining or losing a spouse; losing a job; moving house). **Others:**

- Genetic predisposition
- Faulty learning
- Secondary gain (eg a husband is forced to stay at home with his agoraphobic wife).

Treatment **Symptom control:** Simple listening is a potent way of reducing anxiety. Explain that headaches are not due to a brain tumour, and that palpitations are harmless. Anything you can do to enrich the patient's relationship with others is likely to help.

Cognitive behavioural therapy (p 370) and **relaxation** appear to be the best specific measures, with 50–60% recovering over 6 months. medline:10616949

Behavioural therapy employs **graded exposure** to anxiety-provoking stimuli.

Drugs may augment psychological methods, eg diazepam 5mg/8h po for <6 weeks. Side-effects/withdrawal (p 366) severely limit their usefulness.

NB: **Social anxiety with alcohol abuse syndrome:** Paroxetine helps. medline:11754136

Progressive relaxation therapy Teaching deep breathing using the diaphragm, and tensing and relaxation of muscle groups—eg starting with the toes and working up the body can reduce stress. Practice is essential. Tapes/cds are available for repeated use, to help the learning process. **NB:** in some contexts, eg stress, relaxation is not as good as cognitive restructuring. medline:9554427

Hypnosis This is a powerful mechanism for reducing anxiety. Initially the therapist induces progressively deeper trances using such techniques as guided fantasy and concentration on various bodily sensations, such as breathing. Later, some patients will be able to induce their own trances. It may be very useful, eg medical contexts (eg post-op). medline:9128126

Some remarks on the nature of anxiety

Anxiety is the engine in us, and also our steering wheel, weaving us in and out of the fast or slow lanes of our lives. Some of us deliberately seek out anxiety as a way of feeling alive (think of the tightrope walker, or the falconer who places the meat for his bird between his own eyes). The lives of others are dominated by the imperative of minimizing anxiety, to the extent that some people never leave home, either physically or metaphorically. Anxiety implies heightened awareness, which is why it is one of the dominant colours in the artist's metaphysical palette. The artist makes us anxious so that we see familiar objects differently. Anxiety, up to a point, makes us sit up, and take note; beyond a certain level (different in all of us, and different at different times) anxiety is counter-productive: we become preoccupied with the feeling itself, which chokes our ability to act (*angere*, Latin for to choke, gives us both angina, and anxiety—*OED* 1e vol i 378.) So anxiety is no more a disease or a symptom than the circulation of the blood: it's a disease or symptom: rather it is one aspect of our human condition: it's a part of the way we work. Just as our circulation can fail, so our ways of interacting with anxiety can fail. The most arresting manifestation of this phenomenon is panic (Pan is the god of groundless fear)—ie becoming petrified on being caught in the heart-stopping glare of events beyond our control. The strategies we adopt to recover our equilibrium are revealing: song, humour, camaraderie, and faith in God, gin, or our own genius. When these remedies fail, we turn to the psychiatrist and the strategies of cognitive therapy, hypnosis, and ritual relaxation. When these remedies fail in their turn (not always the case!), we are left on our own: this aloneness is another aspect of the human condition, and out of it grows our creativity and originality, born from the painful union of anxiety, fear, and hope.

Other neurotic disorders

Phobic disorders These involve symptoms of anxiety occurring in specific situations only, and leading to their avoidance. These are labelled according to specific circumstance: agoraphobia (*agora*, Greek for market place) is fear of crowds, travel, or situations away from home; social phobias (where we might be minutely observed, eg small dinner parties); simple phobias, eg to dentists, intercourse, Friday the 13th (triskaidecophobia), spiders (arachnophobia), beetles (paint them red, and put black spots on and they become charming ladybirds). There may also be free-floating 'fear of fear', or fear of disgracing oneself by uncontrollable screaming.

It is important to find out exactly what the phobic stimulus is. It may be very specific, eg travelling by car, not bicycle. Why are some situations avoided? If the patient is deluded that he is being followed or persecuted, paranoia rather than phobia is likely. Panic attacks are lessened by cognitive therapy (p 370) & paroxetine 20–50mg/day PO. medline:10627793 & 10604081

Obsessive-compulsive disorder Compulsions are senseless, repeated rituals. Obsessions are stereotyped, purposeless words, ideas, or phrases that come into the mind. They are perceived by the patient as nonsensical (unlike delusional beliefs), and, although out of character, as originating from themselves (unlike hallucinations or thought insertion). They are often resisted by the patient, but if longstanding, the patient may have given up resisting them. An example of non-verbal compulsive behaviour is the rambler who can never do a long walk because every few paces he wonders if he has really locked the car, and has to return repeatedly to ensure that this has, in fact, been done. Cleaning (eg hand-washing), counting, and dressing rituals are other examples.

Positron emission tomography Positron emission tomography reveals increased blood flow to the orbitofrontal cortex, and reduced blood flow to the caudate nucleus. Successful treatment is reflected by some normalization of metabolism in these areas. medline:7613431

Treatment Behavioural (or cognitive) therapy (p 370). Clomipramine (start with 25mg/day po) and SSRIs (eg fluoxetine, start with 20mg per day po) really can help (even if patients are not depressed): see p 340.

Depersonalization This means an unpleasant state of disturbed perception in which objects (eg parts of the body) are experienced as being changed ('as if made of cotton wool'), becoming unreal, remote, or automatized. There is insight into its subjective nature, and it is not a feature of psychosis—but the patient may think they are going mad. Depersonalization may be primary, or part of another neurosis. Neuroimaging shows that it is associated with functional abnormalities in the sensory cortex in areas where visual, auditory, and somatosensory (cross-modal) data are integrated. medline:11058475

Dissociation (formerly hysteria) *Clinical details: p 334. Example of mass hysteria spread by tv—Pokeman induced 'seizures': see 'the Pokeman contagion'.* n = 12000 medline:11235034

Treatment Behaviour therapy (p 370) is often suitable, provided the patient really wants to change. Antidepressants (p 340) may also help, if combined with behavioural therapy.

Managing stress, post-traumatic stress disorder

'Razing out the written troubles of the brain and razing rooted sorrows'

Signs of stress Insomnia, sleep-walking and other parasomnias (p 390), intrusive thoughts, dyspepsia, chest pain, fainting, palpitations, transitory hypertension, hyperventilation (\pm pins-and-needles), abdominal distension, nausea, irritability, over- or under-eating, depression, poor concentration, poor decision-making, and pre-occupation with trivia.

There are many biological consequences of stress, eg depression; increased weight, BP, and alcohol intake; peptic ulcer; irritable bowel syndrome; delayed wound healing (interleukin-1 β mRNA \downarrow); and migraine.

Relieving stress Smoking, alcohol, and chattering are the most popular methods. If drugs must be given, and there is no asthma, heart failure, or heart block, propranolol (eg 10mg/8h po) may \downarrow autonomic symptoms, but side-effects are common. Some people try safer alternatives, eg exercise, singing, progressive relaxation (p 344), or counselling (p 376).

Post-traumatic stress disorder After great psychological trauma (near-death, shell-shock, rape, crimes of passion, shipwreck) there may be: •Anxiety •Depression •Obsessive recall •Compulsive guilt at one's own survival •Alcohol abuse •Irritability •Bed wetting •School/work refusal. Symptoms may be swift, or take years to come to light.

fMRI implicates the anterior cingulate, with a failure to inhibit amygdala activation \pm \downarrow amygdala threshold to fearful stimuli. The observed hippocampal atrophy is thought to be from chronic hyperarousal symptoms mediated by amygdala activation.

medline:11784878 & 11919308 & 11296313

Treatment may focus on psychotherapeutic reliving of the emotional trauma, with skilled interpretation by the therapist, but success is elusive, and Macbeth's 'sweet oblivious antidote to cleanse the stuffed bosom of that perilous stuff which weighs upon the heart' has yet to be found. After wars the 'talking cures' of psychotherapy have sometimes been found to do more harm than good:* here the best advice may be to try to forget or ignore the past: Macbeth's doctor replied (Act V) that such a patient 'must minister to himself', when he had been unreasonably but royally requested to 'pluck from the memory a rooted sorrow' and 'raze out the written troubles of the brain'—and perhaps tragic literature can offer more than medicine here. *Critical incident stress debriefing* is an unvalidated approach, using principles of proximity, immediacy, expectancy, and simplicity—eg by colleagues or in the format of group discussions of the most troubling aspects of the trauma in question, with ventilation of feelings and normalization of emotional responses.

medline:11232385 & 8829719

There is also evidence that SSRIs and tricyclics can help. cochrane CD002795

Prognosis: Predicting when a given trauma will cause acute, delayed, or chronic post-traumatic stress disorder, or whether it will be a stimulus to growth and have salutary effects on overall functioning (sometimes the case) is impossible. Presence of co-morbid disorders (eg depression) may be important, as well as age at exposure, and social networks.

Prevention: This may involve promoting teamwork, techniques of stress inoculation (by exposure), and desensitization by experience with real casualty management (eg when preparing for war).

www.killology.com

Eating disorders: anorexia & bulimia nervosa

Definition of anorexia nervosa 4 criteria are typically used: medline:11268603

- 1 The person chooses not to eat, leading to dangerous weight loss interfering with normal functioning, eg BMI <17 (BMI = $\text{kg}/\text{height}^2$, in metres), or, more strictly, a weight $<85\%$ of weight expected, taking into account height, sex, and the population the patient belongs to; see p 185.
- 2 Intense fear of becoming obese, even when underweight.
- 3 Disturbance of weight perception (ie feeling fat when thin).
- 4 Amenorrhoea: ≥ 3 consecutive cycles absent (when not on the Pill).

Prevalence $\leq 2.8\%$ at 18yrs; $\leq 1.3\%$ by age 23 (USA figures). medline:11026183

♀:♂ $\approx 20:1$. It is the third commonest chronic illness in teenage girls.

Associations Self-perceived obesity (when BMI is low), depression, excessive exercise, adoption of idols with perfect bodies, low self-worth, lack of parental care, early puberty, excessive alcohol use. medline:7480437

Cognitive behavioural theory. An extreme need to control eating is the central feature of the disorder; in Western societies, a tendency to judge self-worth in terms of shape and weight is superimposed on a paramount need for self-control. medline:9922553

Negative body image ('I hate my body') engendered by western media stereotypes of long-legged beauties are common—but anorexia is much rarer. This fact makes single-factor theories unconvincing. 10% of ♀ siblings may be affected, but this is more likely to show the importance of family behaviour in maintaining symptoms than any direct genetic effect (but concordant pairs of monozygotic twins are described).

Rarely, hypothalamic tumours turn up in those originally diagnosed as having anorexia (amenorrhoea before weight loss is suspicious). Dietary problems in early life, parental preoccupation with food, and family relationships that leave the person without a sense of identity may also be important.

There is scant evidence that the chief problem is psychosexual immaturity; also, antecedent sexual abuse is not a specific risk factor. medline:760925 & 8832119

Symptoms Starts at ~ 16 yrs and may be precipitated by dieting. Prepubertal anorexia does occur: strangely, it is commoner in boys. medline:9989305

The patient attaches excessive importance to weight reduction (an over-valued idea), and she may become deluded that she is repulsively fat when she is, in fact, very malnourished. Excessive exercise, and induced vomiting may occur with abuse of laxatives, diuretics, and appetite suppressants. Many also have episodes of binge eating, followed by remorse, vomiting, and concealment. Suicide attempts and depression often coexist. medline:11026183

Differential diagnosis Depression; Crohn's disease; coeliac disease.

Physical complications of weight reduction Sensitivity to cold, constipation, amenorrhoea, faints, weakness, fatigue, BP \downarrow , K $^+$ \downarrow , glucose \downarrow .

Management What is the urgency? If very weak/faint, K $^+$ \downarrow or glucose \downarrow , then prompt admission, possibly under the Mental Health Act, may be needed for feeding (iv if needed). Otherwise, work on building up a trusting relationship with the patient. Work on several fronts simultaneously: ● Gaining of weight (agreed targets; aim for weight gain of 1.5kg/week, and a final body mass index of 20–25—special foods are not required). ● Treat the medical complications of starvation ● Helping with relationships (eg family therapy) ● Cognitive-behavioural therapy (p 370). medline:7702447 & 9922553

On admission privileges are removed, and restored as weight rises.

Prognosis 2% of anorectic patients die (by starvation), and 16% remain seriously underweight after 4–8yrs. Cardiac complications are the commonest cause of death (K $^+$ \downarrow and prolonged Q-T interval predispose to arrhythmia).

Bulimia

Bulimia is common, affecting ~10% of young women. Diagnostic criteria:

- Recurrent episodes of binge eating
- During binging a lack of feeling of control
- Regular use of mechanisms to overcome the fattening effects of binges (eg vomit-induction, laxatives, excessive exercise)
- Persistent overconcern with body weight
- Body weight higher than required for the diagnosis of anorexia (anorexia trumps the diagnosis of bulimia).

Bulimic patients should be routinely assessed for a concurrent personality disorder using structured clinical interviews—as important associations occur which are likely to reduce the benefits of therapy. medline:10941603

Physical complications: Stomach rupture, haematemesis, and metabolic complications following excessive (eg self-induced) vomiting. There may be painless enlargement of the salivary glands, tetany, and seizures. Russell's sign: calluses form on the back of the hand, following its repeated abrasion against incisors during inducement of vomiting.

Management: Consider behavioural or cognitive techniques (p 368–82). For example, the patient may agree to limit eating to one room in the house, and to eat only at mealtimes. She may agree not to have food in the house, and when shopping, she may agree to buy only those items on a shopping list that she has made when satisfied after a meal. It may help to shop with a friend, and with only sufficient money to buy the items on the list. NB: cognitive-behavioural therapy is better than interpersonal psychotherapy here.

2000; N=220
medline:10807486

Drugs are rarely used, but a small randomized trial shows that fluoxetine (p 341) aids weight control, and reduces craving for carbohydrates—but effects are short-term and a big problem is relapse on stopping the drug.

Organic reactions

Acute organic reactions (Acute confusion, delirium) The key feature is impaired consciousness with onset over hours/days. It is difficult to describe; take any opportunity to be shown it. You have the sense when trying to communicate that he is not with you. He is likely to be disoriented in time (doesn't know day or year) and, with greater impairment, in place. Sometimes he is very quiet/drowsy; sometimes agitated, and you are called when he is disrupting the ward. On other occasions the patient appears deluded (for example, accusing staff of plotting against him/her) or to be hallucinating. If there is no past psychiatric history, and in the setting of a general hospital (ie the patient is physically ill or has had recent surgery); a confusional state is particularly likely if the symptoms are worse at the end of the day.

Differential diagnosis If agitated, consider anxiety (usually readily distinguished on history-taking). If onset uncertain, consider dementia.

Causes Infection; drugs (benzodiazepines, opiates, anticonvulsants, digoxin, L-dopa); $U\&E\uparrow\downarrow$; hypoglycaemia; $P_aO_2\downarrow$; $P_aCO_2\uparrow$; epilepsy; alcohol withdrawal; trauma; surgery (esp. if pre-op $Na^+\downarrow$ or visual or hearing loss). medline:11333420

Investigation $U\&E$, FBC, blood gases, blood glucose, appropriate cultures, LFT, ECG, CXR. Consider LP and CT scan/MRI (p 352).

Management Find the cause. Optimize surroundings and nursing care. Examine with above causes in mind; do tests. Start relevant treatment, eg O_2 .

- 1 If agitated or disruptive, sedation may be needed before examination and tests are possible. Try haloperidol 1–10mg iv/im/po. Use the smallest doses possible (esp. if elderly). Wait 20min to judge effect and need for further dose. In alcohol withdrawal, use diazepam (OHCM p 242). medline:11159573
- 2 Nurse ideally in moderately lit quiet room with same staff in attendance. Repeated reassurance and orientation. A compromise between a quiet room and a place where staff can keep under close surveillance has to be made.

Chronic organic reactions (dementia) Prevalence ~6% of those >65yrs.

Causes and features Worsening memory and global intellectual deterioration without impairment of consciousness. Get a good history from friends/relatives. Exclude depression (may need a drug trial).

- Behaviour: restless; no initiative; repetitive, purposeless activity; sexual disinhibition; social gaffes; shoplifting; rigid routines.
- Speech: syntax errors; aphasia; mutism.
- Thinking: slow, muddled; delusions. Poor memory. No insight.
- Perception: illusions, hallucinations (often visual).
- Mood: irritable, depressed; affect blunt; emotional incontinence (crying++).

Investigating the diagnosis ● Haematology: FBC, B_{12} , red cell folate (macrocytosis suggests alcoholism, or B_{12} or folate deficiency). ESR (malignancy).

- Biochemistry: $U\&E$, LFT, γGT , Ca^{2+} (renal/hepatic failure, alcoholism, malignancy, endocrine causes leading to $Ca^{2+}\downarrow$). T_4 (hypothyroidism).
- Serology: for syphilis (OHCM p 586). After counselling carers, test for HIV if at risk.
- Radiology: CXR (malignancy); CT (hydrocephalus, tumours, subdurals, CNS cysticercosis). As drugs for Alzheimer's become more available, CT becomes more important in ruling out other diagnoses. medline:11237308

Management Exclude treatable illnesses. Explain this to relatives, who may feel unable to look after the immobile, incontinent, aggressive patient. A walking frame, an indwelling catheter, day care, holiday admission, and an attendance allowance can transform this picture. If not, long-stay institutional care may be needed. Consider arranging power of attorney.

Prevention HRT; statins (relative risk 0.29).

medline:11089820

causes of organic reactions

Acute organic reactions:

generative

other CNS

Cerebral tumour or abscess;
subdural haematoma; epilepsy;
acute post-trauma psychosis

Many, eg meningoencephalitis;
septicaemia; cerebral
malaria; trypanosomiasis
Stroke (or TIA); hyperten-
sive encephalopathy; SLE

*U&E imbalance; *hypoxia;
*liver and kidney failure;
non-metastatic cancer; por-
phyria; *alcohol withdrawal
Hyperthyroid or Addisonian
crisis; diabetic pre-coma;
hypoglycaemia;
hypo/hyperparathyroidism
*Alcohol; many drugs (check
Data sheet Compendium);
lead; arsenic; mercury
Thiamine↓; B₁₂↓; folate↓;
nicotinic acid↓

Chronic organic reactions:

*Alzheimer's; *Lewy-body dementia
(eosinophilic intracytoplasmic
neuronal inclusion bodies, p 752)
Creutzfeldt-Jakob; Pick's; (p 754)
or Huntington's diseases (p 750)
Brain tumour; subdural haema-
toma; multiple sclerosis;
Parkinson's; normal pressure
hydrocephalus medline:10773249
Late syphilis; chronic or sub-
acute encephalitis; CNS cysticer-
cosis; cryptococcosis; HIV
Thromboembolic multi-infarct
(arteriosclerotic) dementia; hyper-
tension; anaemia
Liver and kidney failure
non-metastatic or metastatic
cancer

T₄↓; Addison's; hypoglycaemia
hypopituitarism; hypo-/hyper-
parathyroidism medline:10935842

*'Alcoholic dementia'; barbiturate
or bromide abuse; too much man-
ganese or carbon disulfide
Thiamine↓; B₁₂↓; folate↓; nicot-
inic acid↓ (pellagra) medline:9856171

* = leading causes

Example of advice addressed to relatives of demented patients

Alzheimer's disease is usually progressive; with mental functioning getting steadily worse; some problems (eg aggression) may improve over time. Both the rate of change and the length of life vary greatly. Should you try to explain to your relative what the diagnosis is? There is no easy answer. There is no evidence that knowing causes depression (but he may be depressed if he realizes his memory is fading). The advantage of frank talking is that he can participate in his care (so the vexed issue of stopping driving may be easier to handle). Also, during the early stages, he can give consent to future treatments. Most (71%) people would want to be informed if they got Alzheimer's.

Take every opportunity to talk about your predicament with other people in the same position. This is often just as useful as talking to doctors. The Alzheimer's Disease Society exists to put you in touch: uk tel: 020 7306 0606. Accept offers of help, eg with care-giver training programmes, and of daycare for your relative (you certainly deserve, and need, a break from time to time). Lock up any rooms in the house which you do not use. Your relative will not notice this restriction—and this may make your life much easier.

Lock drawers which contain important papers or easily-spoiled items to prevent him storing inappropriate things in them, such as compost, or worse. Remove locks from the lavatory—so he/she cannot get locked in.

Sexual activities may stop. Spouses should try not to fall into the trap of asking 'What's the matter with me?' medroxyprogesterone acetate 100–200mg/2 weeks IM may help if hypersexuality becomes a problem. medline 2957362

Prepare yourself psychologically for the day when he/she no longer recognizes you. This can be a great blow, unless you prepare for it.

In the UK, apply to Social Services for Attendance Allowance + council tax rebate.

Alzheimer's dementia (AD)

This is the major neuropsychiatric disorder of our times, dominating not just psychogeriatric wards, but the lives of many thousand sons, daughters and spouses who have given up work, friends, and all their accustomed ways of life to support relatives through the last long years. The struggle of caring for loved ones through terminal illness always puts us on our mettle never more so than when that loved one's personality disintegrates, and the person who is loved is gone long before their eventual death. *Suspected Alzheimer's disease* in any enduring, acquired deficit of memory and cognition, eg as revealed in the mental test score and other psychometric tests (p 353). Onset may be from 40yrs (earlier in Down's syndrome)—so the notions of 'senile' and 'pre-senile' dementia are blurred (and irrelevant). Chemically, there is a deficit of neurotransmitter acetylcholine.

Genetics This is complex (defective genes on chromosomes 1, 14, 19, 21) the apoE4 variant is a major risk-factor, also bringing forward age of onset).

Diagnosis *Exclusion of other causes* (OPPOSITE) show dementia is present; *biochemistry* etc. (p 350) rule out hypothyroidism, $\downarrow B_{12}$, etc, but *histology* (plaques + neurofibrillary tangles + cortical β -amyloid protein) is needed to tell apart vascular, frontal lobe, Lewy-body (p 752), and Pick's dementias (p 754). In the elderly, mixed dementias are very common, further complicating things. *Imaging* is nonspecific, although serial MRIs (separated by >3 months) may show \uparrow loss in the medial temporal lobe (especially the hippocampus). *Blood flow images* typically show hypoperfusion in the temporal-parietal areas, and more anteriorly in frontotemporal dementia. *Advanced genotyping* (e4 allele) may help, as may *CSF analysis* (β -amyloid \downarrow ; tau protein \uparrow). Lancet 2002 359 1530.

Presentation In stage I of AD there is failing memory and spatial disorientation. In stage II (which follows after some years) personality disintegrates (eg with increased aggression and focal parietal signs, eg dysphasia, apraxia, agnosia, and acalculia). Parkinsonism may occur. She may use her mouth to examine objects (hyperorality). In stage III she is apathetic (or ceaselessly active—akathisia), wasted, bedridden, and incontinent, \pm fits and spasticity.

Mean survival 7yrs from onset.

Potential strategies Examples include augmenting nerve growth factor which is taken up at nerve endings, and promotes nerve cell repair. Also:

- Preventing the breakdown of acetylcholine, eg donepezil, below.
- Stimulating nicotinic receptors, which may protect nerve cells.
- Inhibition of the enzymes that snip out β -amyloid peptide from APP, (amyloid precursor protein), so preventing fibrils and plaques.
- NSAIDs may stop microglial cells secreting neurotoxic cytokines and glutamate and so \downarrow APP formation. Using NSAIDs for >2yrs \downarrow AD risk by 80%. medline:1179421
- Regulation of calcium entry (mediates the damage of neurotoxins).
- Preventing oxidative damage by free radicals, ie a possible rôle for HRT and vitamins: AD is associated with \downarrow plasma folate & \uparrow homocysteine. medline:1184484

Liaise with an expert about increasing CNS acetylcholine by inhibiting the enzyme causing its breakdown (donepezil; rivastigmine). Antioxidants, eg ginkgo biloba (EGB 761, 120mg/day po) are endorsed by randomized trials. EGB may delay need for institutional care, but not necessarily its duration. medline:1180323

Practical issues \blacktriangleright See *Help for relatives of the demented*, p 351. Exclude treatable dementias (B_{12} , folate, syphilis serology, T_4 , ?HIV). Treat concurrent illnesses (they worsen dementia). In most, the dementia progresses. Involve relatives and relevant agencies.

Prognosis Vitamins \pm HRT (as above); cognitively stimulating hobbies help. A 1-point rise in cognitive activity score can \downarrow risk of AD by 33%. medline:1185154

Mental test score

This is a quantifiable, standardized way of measuring someone's mental functioning. It is most useful for serial measurements. Maximum score 34; normal ≥ 29 ; a change of ≥ 5 is meaningful. See also the *Mini-Mental State Examination*, *OHCM*, p 535.

Knows his name	score 0 or 1	Recognizes 2 people	
Knows his age	score 0 or 1	score 0, 1 or 2	
Time	score 0 or 1	Birthday	0 or 1
(to nearest hour is ok)		Town of birth	0 or 1
Teach name and		School	0 or 1
address: test recall after 5min:		Former occupation	0 or 1
Mr John Brown	score 0, 1 or 2	Name wife, sibling or	
42 West Street	score 0, 1 or 2	next of kin	0 or 1
Gateshead	score 0, 1 or 2	Monarch's name	0 or 1
Day of the week	score 0 or 1	Prime minister's or head	
Date of month	score 0 or 1	of state's name	0 or 1
Current year	score 0 or 1	Give years of:	
		World War I	0 or 1
Place: eg hospital	score 0 or 1	World War II	0 or 1
name of hospital	score 0 or 1	Months of year	
name of ward	score 0 or 1	backwards	0, 1 or 2
name of town	score 0 or 1	Count 1-20	0, 1 or 2
		Count 20-1	0, 1 or 2

Other possibly significant questions Increasingly, do you find that:

- You forget what you are saying or reading in mid-sentence?
- You have to rely on lists whereas previously this was not necessary?
- Thought is slow or imprecise, harmonizing poorly with motor control?
- Mental agility is lacking, with powers of concentration declining?
- Are there difficulties executing fast movements of eyes or limbs, or difficulties in walking?, eg with spastic ataxic gait or quadriparesis of *HIV-1-associated cognitive/motor complex*, or psychomotor retardation, \pm release reflexes such as a snout response, or hyperactive deep tendon reflexes.

IB: Including an 'informant report questionnaire' improves the efficiency of the mental test score as a screening tool for dementia. medline:9812113

Psychosis (including mania)

► Beware labelling people; remember that even during the best of times only a thin veil separates us from insanity. In its most florid form, psychosis is the archetype of the layman's 'madness', and it signifies a state of mind in which contact with reality is lost and the landmarks of our normal mental processes are suspended and turned awry, with an abandoning of constraint imposed by reason and morality. But, the *usual* picture is much less obvious: the patient may be sitting alone, quietly attending to his or her voices.

Key features suggesting psychosis are: hallucinations, delusions, and thought disorder—defined on p 316 & p 358. If one of these features is present, the diagnosis is limited to 4 entities: schizophrenia (p 358), a disorder of affect (ie mania or depression, p 336), a paranoid state, or an organic, ie physical disease. So the term psychosis is not in itself a diagnosis, but is a useful term to employ, while the underlying diagnosis is being formulated.

Mania and hypomania We normally adapt our pace to the obstacles we encounter—but in mania we march a round in circles, like a tornado picking up internal speed as we go, with the potential to destroy everything beneath the conquering tread of an unfettered will, while exhibiting these signs:

- Euphoria • Hyperactivity • Hallucinations • Over-assertiveness
- Appetite ↑ • Pressure of speech • Disinhibition • Grandiose delusion
- Insight ↓ • Sexual desire ↑ • Spending sprees • Self-important ideas

Less severe states are termed *hypomania*. If depression alternates with manic features, the term *manic depressive psychosis* is used—particularly if there is a history of this in the close family. Cyclical mood swings without the more florid features (listed above) are termed *cyclothymia*.

Treatment: Sedation may be needed (chlorpromazine 50mg/8h po; droperidol 5–15mg/4h iv). Zuclopenthixol acetate, 50–100mg im into a buttock is an alternative (perhaps⁶), if you know that the patient can tolerate neuroleptics; it lasts 2–3 days. *Prophylaxis* (eg if more than one episode of mania).

If compliance is good, and U&E, ECG, and T₄ normal, give lithium carbonate 125mg–1g/12h po as follows. (Note: intracellular levels take time to build up.)

- Adjust the dose to achieve a plasma level of ~0.7–1.0mmol/L Li⁺, measured on the 4th–7th day of treatment, 12h (± h) after the preceding dose.
- Check Li⁺ levels weekly until the dose has been constant for 4 weeks then monthly for 6 months, and then every 3 months, if stable. Do more often if on diuretic, or a low salt diet or pregnant (avoid Li⁺ if possible).
- If levels are progressively rising, suspect progressive nephrotoxicity.
- Do plasma creatinine and TSH every 6 months; Li⁺ affects thyroid and kidney (hypothyroidism; nephrogenic diabetes insipidus).
- Avoid changing proprietary brands, as Li⁺ bioavailability varies.
- Make sure you know how to contact the patient by phone if Li⁺ levels are high (>1.4mmol/L prompts urgent contact with patient to adjust dose).
- Toxic effects: blurred vision, D&V, K⁺ ↓, drowsiness, ataxia, coarse tremor, dysarthria, hyperextension, seizures, psychosis, coma, and shock.

Organic psychoses These follow some physical stimulus: narcotics; amphetamines; cocaine; lysergic acid; alcohol; glue or solvent abuse; CNS trauma or tumour; epilepsy; puerperal psychosis (see p 148, the stimulus is birth). Associated stress, life events, and tiredness are also important.

Paranoid states This implies paranoid symptoms (delusions or hallucinations) which concern a patient's relationships with others, and in whom using one of the other 4 categories of psychosis (above) is unjustified. A common form is a persecutory delusion ('there is a conspiracy against me'). Morbid jealousy (the Othello syndrome—p 754) is a rarer example.

Options when lithium does not give good control

Lithium is inadequate for the many people with bipolar illness (especially if there is rapid cycling), even if supplemented by antidepressants (below) and neuroleptics. Semisodium valproate and carbamazepine (OHCM p 366; 100mg/8h po) are next steps. *Dose example for semisodium valproate* (there is wide variation): 250mg/8h po, increased quite rapidly to the most effective dose, eg 1g/12h (swallow whole; do not chew). medline:10770458

Some authorities say that semisodium valproate is better than lithium, but the evidence is equivocal.*

Even with these agents, patients often need additional drugs, eg nimodipine (and related dihydropyridine calcium channel blockers), lamotrigine, gabapentin, topiramate, and atypical neuroleptics (p 360). medline:11252642

Preferred antidepressants Bupropion, selective serotonin reuptake inhibitors (SSRIs), and venlafaxine are said by expert committees to be best. Taper from 2 to 6 months after remission. medline:10895797

Monoamine oxidase inhibitors should be considered for patients with anergic (=lacking in energy) bipolar depression. medline:10826663

ECT (p 342) may be very helpful. In one small randomized study, ECT patients improved more during the 1st 8 weeks than did patients given lithium. ECT was especially helpful with mixed symptoms of mania and depression and/or extreme manic behaviour. After 8 weeks there were no differences between the groups. N=14
medline:2899425

A more subtle approach is teaching about early signs of relapse, and benefits of early presentation. This decreases time to 1st manic relapse, and improves social functioning and employment prospects. N=6
medline~9888904

Cognitive and family therapies also help. Also, teach about social rhythm therapy (no stress; regular meals & bedtimes, etc). medline:11153813 & 11256459

NB: Not everyone will need Li⁺ prophylaxis after one episode of mania. Numbers needed to treat to prevent one relapse ≈ 3.

T Dinan 2002 *BMJ* i 990 and K Macritchie 2001 *Cochrane CD* 003196—but there may be a special rôle for semisodium valproate in rapid cyclers.

Schizophrenia: history & current concepts*

Schizophrenia is a common mental disorder which typically presents in adulthood with delusions (p 316), hallucinations, and disordered thought (positive symptoms). There is also a more chronic picture with withdrawal, apathy, emotional blunting, and slowness of thought (negative symptoms). *Incidence* ~0.15/1000/yr. *Prevalence* ~3/1000. It is a disorder with major implications, for the patient, his or her family, and it forms the basis for much of the chronic workload of psychiatrists and psychiatric nurses.

Note that models of schizophrenia are not necessarily inconsistent with each other. Stress for example, has both social and biological aspects, and is important in the genesis of schizophrenia: stress at work, stress in the family, urban stress, and stress associated with immigration.

Older models *Behavioural disorder* is a pattern of well-learned avoidance responses. The behaviour is the illness. *As a family disorder* Here a 'schizophrenogenic' mother gives conflicting messages (the 'double bind') eg to her daughter who is having difficulty with her exams: 'Your exams are very trying, and I do so understand why you have locked yourself in your room. I'll do anything I can to help. You know you are putting me and dad under a lot of stress: if this leads us to separate, it will be your fault.' NB: most mothers are very supportive of their schizophrenic offspring: don't alienate them!

Newer models of schizophrenia Schizophrenia is now often thought of as a *physical/chemical disorder*. PET images show subtle changes affecting the hippocampus and temporal lobes (where glutamate receptor subtypes are reported to be missing) in schizophrenia. The cerebral ventricles are also enlarged. Concepts derived from cognitive neuroanatomy are increasingly employed to throw light on schizophrenia. There is evidence that:

- Schizophrenic patients with poverty of speech show low blood flow in the dorsolateral prefrontal cortex (DLPFC) on the left. *medline:1433066 & 909350*
- During hallucinations muscle activity resembling phonation is detectable in the tongue. Pre-vocal speech can be made audible by microphones, and this may correspond to what the hallucinations are telling the patient to do.
- We are all prone to paralysis of the will (contrary to the views of the soldiers in their pre-psychotic phases, such as Othello, for whom 'to . . . be in doubt is [at] once to be resolved'.) When psychosis arrives we can expect to find disconnections in pathways of willed action: imaging during tasks involving choice (as opposed to the same manual task but pre-specified) shows these pathways connect the anterior cingulate cortex, the basal ganglia, the dorsolateral prefrontal cortex (DLPFC), and the supplementary motor area.
- In schizophrenic patients there is an abnormal lack of connectivity between DLPFC and the superior temporal cortex.

Animal models of schizophrenia Cannabinoid receptor gene knockout mice show behavioural alterations paralleling schizophrenia. *medline:1139911*

One conclusion from this and other evidence is that we should regard schizophrenia as a white-matter neurodevelopmental encephalopathy affecting the interconnections of the associative centres of the brain (if there is no distinct focal lesion). The initial event may be intrauterine stresses such as poor nutrition or infection. Frith has postulated that these events lead to a *defect in self-monitoring* such that the patient does not appreciate that he or she has willed inner speech, and these pre-vocal words therefore take on the characteristics of hallucinations. *medline:765100* Negative features arise from failure of the will due to events (or lack of events) in the dopaminergic pathways of willed action described above.

*Seminal paper: Postpsychiatry: P Bracken 2001 *BMJ* i 724 www.bmj.com/cgi/content/full/322/7288/724

Testing neurodevelopmental models

To untrained observers, a young man's first schizophrenic episode seems like a bolt from the blue: unheralded, and cataclysmic. But we know that there are subtle but telling differences in early behaviour of those going on to get schizophrenia, eg there is excess solitary play ($p < 0.05$; $N = 5362$) and walking is delayed by a few months ($p < 0.005$). medline:7968076

Schizophrenia is also associated with early rupture of membranes, gestational age < 37 wks and incubator use (?reflecting hypoxia). medline:10478777

Poor social functioning, poor organisational ability, and low intellectual functioning in boys predict schizophrenia 4–10 years later. medline:10484941

Twin studies In monozygotic twins discordant for schizophrenia, the twin with the smaller temporal lobes and larger ventricles has the schizophrenia (but twin studies are fraught with bias medline:11780600). medline:10023939

One abiding difficulty with studying the behaviour and CNS characteristics of those with chronic schizophrenia is that it is impossible to tell for sure whether the events observed or the structural differences measured are the cause or the effect of the schizophrenia. Suffering family breakdown or living in an institution, or taking neuroleptic medication are themselves highly significant events.

Studies of first-episode patients Some of these confounding events can be obviated by imaging the brains of those undergoing their first-ever schizophrenic event. Such studies show reduced brain volumes (\downarrow by 3%), along with more specific deficits in medial temporal lobe structures such as the amygdala (known to be important in the recognition and expression of emotion) and hippocampus (needed for memory function). Emotion and memory are affected in schizophrenia, and the volume of these areas is reduced, typically by $\sim 10\%$. The thalamus is also abnormal, and may be the source of hallucinatory events.

No-episode-yet 'patients' If neurodevelopmental models of schizophrenia are correct, then it is predicted that individuals who have not yet had their first episode will nevertheless have structural brain anomalies. We now have evidence that this is the case. medline:10023948

The chief known risk factor is family history (risk \uparrow by 40-fold if a close relative is affected, but no strongly linked genes have been found). On MRI, these people (aged 16–25) have anomalies similar to 1st-episode patients.

So, in this sense, schizophrenia is becoming a little more predictable, and, in time, this may pave the way for early detection and novel treatments.

The postpsychiatry viewpoint

www.bmj.com/cgi/content/full/322/7288/724

The above views reflect the dominant CNS-based model of schizophrenia. But we are evolving a counter 'postpsychiatry' paradigm which centres not on neurochemical events but on social and cultural context. In this view, a psychotic idea is interpreted as a valid experience with social, cultural, and personal meaning. The psychiatrist Marius Romme, for example, has established worldwide networks of voice-hearers who interpret their intrapsychic phenomena in creative ways. www.hearing-voices.org.uk/

Psychiatrists are paying ever closer attention to these ideas because of the universal trend towards client-centred approaches, and an increasing recognition that science and reductionist analysis can only ever provide part of the picture—which is really one of interdependence between biological concepts of disease and social conceptions of health. medline:8184373

Schizophrenia: *diagnosis and prognosis*

Schneider's 1st-rank symptoms Seen in $\leq 70\%$ of patients.

- *Thought insertion*: 'Someone is putting thoughts into my head.'
- *Thought broadcasting*: 'People hear my thoughts as they occur.'
- *Thought withdrawal*: 'Thoughts are being taken out of my head.' (Thoughts may also stop in midflow, resuming on a different tack.)
- *Passivity*: 'Someone's controlling my thoughts and movements; I'm a robot.'
- Hearing 'voices commenting on my actions' or referring to the patient in the third person (or the repeating of thoughts—*écho des pensées*).
- Primary delusions, in addition to the thought-delusions above.
- Somatic hallucinations, ie the experience of being the passive and reluctant recipient of bodily sensations imposed by some external agency—eg the experience of being sexually penetrated.

Other hallucinations/delusions occur in depression, mania, and organic reactions. Having one of Schneider's first rank symptoms is very suggestive of psychosis, but doesn't help diagnose schizophrenia specifically. Diagnostic systems for schizophrenia that are heavily based on first rank symptoms may arise from a tautological definition of the disorder. medline:1044845

Cognitive and negative symptoms are also important, medline:1153385 eg cold, unemotional, unempathic, 'as if a pane of glass separated me from others'; incongruity of affect, eg laughing at bad news; and *knight's move thoughts* that change direction, flying off at tangents, with odd logic: 'his ideas seemed to move on a deeper level, like roots spreading below a layer of compost a code uncracked which waits for a clue.' Graham Greene Under the Gard

With psychotic features and *no* first rank symptoms, look meticulously for depression or mania. Ask yourself: is the content of the delusion or hallucination suggestive of either depression or mania (guilty or grandiose)?

Cannabis, amphetamines, and other drugs can cause schizophrenia-like symptoms. medline:11278156 A good history, and understanding the vocabulary of drug abusers (*OHCM* p 552–3) will usually point you in the right direction

The most frequent symptoms:

Lack of insight	97%
Auditory hallucinations	74%
Ideas of reference (p 316)	70%
Suspiciousness	66%
Flatness of affect	66%
Voices speaking to patient	65%
Delusions of persecution	62%
Thought-delusions	~50%

The most frequent behaviour:

Social withdrawal	74%
Underactivity	56%
Lack of conversation	54%
Few leisure interests	50%
Slowness	48%
Overactivity	41%
Neglect of appearance	30%
Odd postures or movements	25%

Best prognosis if: Sudden onset; no negative symptoms; supportive home. Also: high scores on measures of expressed emotion, premorbid psychosocial evolution, female sex, and -ve family history (data from *WHO's disability assessment schedule* medline:7781589). Overall, only 10% ever have one episode. With treatment, $\leq 7\%$ stay in hospital for >2 yrs after first admission. 28% go at least 2 years without needing further hospital admission. The lifetime risk of schizophrenia in the general population is $\sim 1\%$.

The family Get expert help. Create agreed care plans, specifying all options. Is housing adequate? Monitor family stress levels (plan what to do if levels rise). Family teaching on schizophrenia and problem-solving skills reduces relapses, hospitalizations, and helps drug compliance. medline:1061662

Schizophrenia: management

Aims These are to treat the patient and the illness as well as possible, and to provide support and advice to the relatives. Yes, if the patient is a danger to himself or others, or hospital is the only place where you can be sure the patient will get the treatment he needs. Treat acute symptoms with an antipsychotic agent. The following is based on www.nice.org.uk advice (2002). Risperidone (and quetiapine, zotepine, etc.) have fewer s_{es} than older neuroleptics, and work better (they combine serotonin and dopamine receptor antagonism). medline:11873701

All antipsychotics have s_{es} (below), so discuss with the patient or, if impossible, with someone close to them. Antipsychotics are only one part of any care-plan. medline:12046641 & 12047021
2mg po on day 1; 4mg on day 2; slowly increased to 6mg/24h if needed (max dose: 5–8mg/12h, exceptionally). If elderly, start with 0.5mg/12h and work up slowly by 0.5mg/12h increments to 2mg/12h. medline:12046641 & 12047021 Insomnia, agitation, anxiety, headache, drowsiness, concentration↓, constipation, abdominal pain, hyperprolactinaemia, amenorrhoea, gynaecomastia, priapism, incontinence, pulse↑, BP↑, rhinitis, oedema, poor T° regulation, glucose↑, ketoacidosis, wcc↓.

In some parts of the world only cheap antipsychotics are available, eg chlorpromazine 25mg/8h po, up to 1g/day (500mg if elderly)—may cause extrapyramidal effects, akathisia, photosensitivity, jaundice, and other effects.*

After a few months, antipsychotics reach full effect. Longer-term management includes helping the patient engage in as normal a life as possible, in a planned way. Aim to reduce relapse risk by continuing medication, and trying to ensure a stress-free environment. Relatives need help to be neither critical nor overprotective. Cognitive therapy (p 370) ↓psychotic symptoms. Time is devoted to systematically reviewing the start of symptoms. Patients are questioned in detail to reveal faulty reasoning, and then encouraged to monitor psychotic symptoms, and develop coping strategies, eg: •Changing the focus of attention •Relaxation techniques •Modifying exacerbating behaviours. The use of these have been confirmed in trials, and their effects may be more humanizing than the ready resort to the 'liquid cosh' (below)

Psychotic violent patients 150 people have been killed in the uk in 1991–7

- Recognize early warning signs: tachypnoea, clenched fists, shouting, chanting, restlessness, repetitive movements, pacing, gesticulations. Your own intuition may be helpful here. At the first hint of violence, get help. If alone, make sure you are nearer the door than the patient.
- Do not be alone with the patient; summon the police if needed.
- Try calming and talking with the patient. Do not touch him. Use your body language to reassure (sitting back, open palms, attentive).
- Get his consent. If he does not consent to treatment, emergency treatment can still be given to save life, or serious deterioration.
- Use minimum force to achieve his welfare (but may entail 6 strong men).
- If one of the following IM drugs is needed, monitor BP every 4h.
- Later, liaise with colleagues about long-term care. Will the outside world be at risk if community care is used? If so (or if the person is at danger from suicide or self-neglect) use the Care Programme Approach (CPA, p 399) with a key worker (eg a community psychiatric nurse) trained in risk-management and assessment, who can co-ordinate multidisciplinary review meetings to monitor progress. These requirements may be counter-productive, if no extra funds are available and the key worker's chief aim is to keep people out of hospital.
- Haloperidol: 2–30mg IM, then 5mg/1h, max total 60mg; switch to an oral agent as soon as possible.*

If no response to risperidone Try eg olanzapine. medline:11875967 &: 11835695

If no response to olanzapine Clozapine is licensed in this context. Specialist monitoring for early detection of serious neutropenia** is required. (Olanzapine can also cause agranulocytosis, but only 12 cases are known.) Additional sEs: seizures, drowsiness, salivation↑, LFTs↑. medline:11823268

IB: plasma levels of clozapine and olanzapine are lower in smokers than in nonsmokers via induction of cytochrome P4501A2. Quitting tobacco or quitting cannabis can cause toxic levels. medline:11981356

Children and adolescents Because of their efficacy, newer antipsychotics are coming to the fore in child psychiatry; get specialized help if seizure threshold is low. ►Monitor growth, body weight, and glycaemic status. Expect dose changes if smoking starts or stops. medline:11772153

'Second-chance' programmes If stable on traditional neuroleptics, but not mentally well, should a switch be made to a new antipsychotic? In one study, 43% switched to risperidone were discharged to supervised residences on the basis of their clinical improvement. medline:12045315 &: 11910255

Older agents are more likely to cause: ●Parkinsonism ●Akathisia (restlessness—may be very distressing; so use lowest possible dose—treatment may be needed with propranolol 20mg/8h po ± cyproheptadine 4mg/6h po). medline 11763011 ●Dystonic reactions (torticollis, opisthotonus), often in 1st weeks of treatment, and responding to biperiden 2mg po up to 4x daily. ●Tardive dyskinesia (chewing, grimaces, choreoathetosis) may be irreversible; but try tetrabenazine 12.5–50mg/6–24h po (eg after waiting to see what happens for ~6 months after stopping the drug). Neuroleptic poisoning: OHCM p 829.

IB: risperidone, olanzapine, and ziprasidone show dose-related extra-pyramidal effects, but clozapine and quetiapine apparently do not. Quetiapine does not require the regular blood monitoring mandated for clozapine. medline:10908478 &: 12046640

Older neuroleptics may be less likely to help negative features such as withdrawal.

►Try to prevent wcc↓ by: ●Only starting clozapine in hospital. ●Explain that any symptoms of infection must be reported immediately. ●Taper off conventional neuroleptic ●Monitor wcc and differential every week for first 18 weeks, then every fortnight if ok. ●If all is well for 1 yr, monitor monthly; and for 4 weeks after stopping. ●Avoid co-therapy with wcc-lowering drugs. ●If wcc <3 × 10⁹/L (or neutrophils <1.5) at any time, stop clozapine permanently.

Drug problems and addiction (► See OHCM p 552)

Definitions

A drug's early effects are later achievable only by using higher doses. *Withdrawal symptoms* The effects experienced when a tolerance-inducing drug is withdrawn; these may be psychological (restlessness and craving when the drug is withdrawn) or physical, such as the 'cold turkey' which occurs in narcotic withdrawal—dilated pupils, D&V, tachycardia, sweating, cramps, and piloerection. *Drug maintenance* Only continued doses prevent withdrawal symptoms (compulsive activity to avoid this is *drug seeking*). The drugs involved are the opiate derivatives, amphetamine, cocaine (p 550), lysergic acid, hydrocarbons ('glue sniffing'), barbiturates and, to a lesser extent, cannabis. Opiate withdrawal may be eased by methadone, eg 20–70mg/12h po, reducing by 20% every 2 days, ideally as part of a regimen in which a contract is made with the patient (p 339), eg in a specialist clinic or in primary care, provided the GP has an interest and ongoing commitment. In the UK, 40% of prescriptions for methadone are given by GPs. But most GPs are unwilling to become too closely involved for fear of attracting too many addicts, who might end up abusing them, and their staff with the added risk of their methadone ending up on the black market.

A non-addictive alternative is lofexidine (α_2 -noradrenergic agonist like clonidine)—eg 0.2mg/24h po, increased by 0.2–0.4mg increments/day (max 2.4mg/day); a 5-day régime may be better than 10-day ones. *SE*: drowsiness, BP↓, pulse↓, dry mouth, rebound hypertension on withdrawal. medline:10598213

Cocaine use by patients on methadone is a big problem, and is associated with a poorer prognosis. Disulfiram has a rôle here. medline:10723858

Narcotic abusers must, by UK law, be notified to the Home Office. Barbiturate withdrawal may cause seizures and death, and withdrawal should be as an inpatient, giving one-third of the previous daily dose as phenobarbital. Lower the dose over 14 days. For nicotine addiction, see p 452.

Psychological support *Cognitive therapy* (p 382) helps adults and adolescents and is a cost-effective adjunct to methadone. medline:9283297

Cognitive behavioural therapy (p 370) is known to increase abstinence in urban crack cocaine abusers. N=138
medline:980371

When to suspect drug addiction • Convictions for crime, to buy drugs

- Any odd behaviour, eg with visual hallucinations, elation or mania.
- Unexplained nasal discharge (cocaine sniffing, p 550).
- The results of injections: marked veins; abscesses; hepatitis; AIDS.
- Repeated requests for pain killers—with only opiates being acceptable.

Counselling parents about how they can counsel their children

- If possible, father and mother should agree on a plan together, along with someone who knows the child (eg teacher or GP).
- Accept the child; and try not to seem authoritarian or bossy.
- Find out what the child's attitudes are to drugs. He may think that hard drugs are no more harmful than alcohol and tobacco.
- Talk about drugs with siblings, showing them cool ways of saying 'No.'
- Confidential phone advice (in the UK 0800 515927).

Dangers of HIV infection (OHCM p 580) if drugs are injected alter the approach to long-term management. It is important to try to maintain a relationship between the health team and the addict, and to educate the patient about avoiding high-risk behaviour. There is controversy over the extent to which addicts should be supplied with clean needles and controlled doses of addictive drugs. See OHCM p 552–3.

Causes *Inherited vulnerability* is as important as *social factors*. medline:992604

Alcohol-related problems

Repeated drinking harms a person's work or social life, and causes ~75,000 years of life lost prematurely, each year (UK). Addiction implies:

Difficulty or failure of abstinence Often aware of compulsion to drink
Narrowing of drinking repertoire Priority is to maintain alcohol intake
Increased tolerance to alcohol Sweats, nausea, or tremor on withdrawal

Ask about tolerance, worry about drinking, 'eye opener' drinks used in the mornings, amnesia from alcohol use, and attempts to cut down. 2 points is WEAK +ve (?more sensitive than CAGE questions). medline:9669791 & 10487741

Alcohol and organ damage Liver (normal in 50% of alcoholics). Fatty liver: acute, reversible; hepatitis; 80% progress to cirrhosis (hepatic failure in 10%); cirrhosis: 5yr survival 48% if alcohol intake continues (if it stops, 77%).

Signs: Poor memory/cognition—may respond to multiple high-potency vitamins IM; cortical/cerebellar atrophy; retrobulbar neuropathy; fits; falls; accidents; neuropathy; Korsakoff's/Wernicke's encephalopathy (OHCM p 736).

GI: D&V; peptic ulcer; erosions; varices; pancreatitis. **Marrow** Hb↓; mcv↑.

Heart: Arrhythmias; cardiomyopathy; fewer MIs, ie beneficial (only if >55yrs).

Alcohol drug levels Regular heavy drinking induces hepatic enzymes, but drinking inhibits enzymes. (Do not indulge in both and hope for the best). Be alert with phenytoin, warfarin, and tolbutamide, etc. NB: paracetamol may cause ↑N-acetyl-p-benzoquinoneimine (it is hepatotoxic).

Withdrawal signs (Delirium tremens) Pulse↑; BP↓; tremor; fits; visual or tactile hallucinations, eg of animals crawling all over one. **Treat** diazepam

Admit; monitor vital signs (beware BP↓). ●For the first 3 days give diazepam generously, eg 10mg/6h PO or PR if vomiting—or IVI during fits, Chlordiazepoxide is an alternative. After the first days, reduce diazepam (eg 10mg/8h PO from day 4–6, then 5mg/12h PO for 2 more days). β-blockers, clonidine, carbamazepine, and neuroleptics (if no liver damage) may be used as adjunctive therapy but are not recommended as monotherapy. medline:9214531

Treatment Key determinants of success are the patient's commitment and willingness to comply with treatment—so it is worth taking time to explore issues of concordance between your own ideas and those of your patient.

Treat any co-existing depression (p 336–8). Refer to a specialist service. Group psychotherapy in self-help organizations such as Alcoholics Anonymous may help, ± agents which produce an unpleasant reaction if alcohol is taken (eg disulfiram 800mg stat then ↓ to 100–200mg/24h PO over 5 days). Reducing the pleasure that alcohol brings (and craving when it is withdrawn) with naltrexone 25–50mg/24h PO (an opioid receptor antagonist used in opiate addictions) can halve relapse rates. medline:11784454 SE: vomiting, drowsiness, dizziness, cramps, joint pain. CI: hepatitis; liver failure. Liaise with experts on its best use. Acamprosate (OHCM p 325) can treble abstinence rates. CI: pregnancy, severe liver failure, creatinine >120μmol/L; SE: D&V, weight fluctuation; dose example: 666mg/8h PO if >60kg & <65yrs old. Economic analysis supports its use in some communities. medline:10899732

Antihemorrhagic agents Disulfiram, naltrexone, and acamprosate are not recommended for the treatment of alcohol dependence. Education, counselling, goal-setting + monitoring of γGT in those who have social or physical problems from alcohol, but who do not exhibit full dependency.) 4 out of 8 randomized trials have shown that γGT falls in the intervention group, but none has shown convincing improvement in alcohol-related morbidity. More costly, specialized interventions fare no better.

Homelessness is common; help with housing, problem solving, communication, drink refusal, & goal setting can help this desperate problem. medline:9642893

Prevention See p 454.

Personality disorders and psychopathy

Personality comprises lasting characteristics which make us who we are: easygoing or anxious; optimistic or pessimistic; placid or histrionic; ambitious or stay-at-home; fearless or timid; self-deprecatory or narcissistic (self-love, founded on a grandiose belief in one's unique superiority). medline:11345846

Personality can change and develop, and it may even change quickly, eg after religious conversion in which a timid man is remoulded into a fearless activist. Personality is a spectrum lying between the above opposites. Statistical analysis reveals that all these distinctions may in fact overlap, and are describable in terms of a few orthogonal dimensions (eg neuroticism/psychoticism; introvert/extrovert). Those with abnormal personalities may then be defined as occupying the extremes of the spectrum. Abnormal personality only matters if it is maladaptive, causing suffering either to its possessor or his associates. In general, psychological symptoms which are part of a personality disorder are harder to treat than those arising from other causes.

Psychopathy 'He dislikes showing his feelings, and he'd rather be cruel than put his real feelings into words... he doesn't care for anyone and perhaps never will.' So says Dostoyevsky; lesser psychologists dwell* on reckless, antisocial acts, impulsivity, lack of guilt ± nonconformity with social and legal codes. Dostoyevsky's definition lasts because of its brevity and because of that telling word *perhaps*. Can we change? What must change before psychopaths can love? This little *perhaps* blowing to us on a wind from 19th-century Russia sends a shiver down our 21st-century spines: perhaps all these psychopaths we lock up *might* be able to change. What needs to be unlocked? Read *Crime and Punishment* to find out. www.bartleby.com/318/32.html#39 Psychotherapists are beginning to work in this area, not always successfully. It may be easier to help the old rather than the young. medline:10028212 & 10484939

Causation: Brain damage, genetic and cultural inheritance, parental psychiatric illness or laxity have been suggested, but none is pre-eminent.

Treatment is problematic as patients are seldom in a frame of mind that is likely to promote change. Peer group pressure (as may occur in group therapy, p 372) may be a motivating force. It is rarely wise to use drugs, but there is evidence that SSRIs (p 341) may be beneficial in aggressive personality disorders.

Other personalities **Obsessive personality** The rigid, obstinate bigot who is preoccupied with unimportant (or vital) details.

Histrionic personality The self-centred, sexually provocative (but frigid) person who enjoys (but does not feel) angry scenes.

Schizoid personality Cold, aloof, introspective, misanthropic. medline:11019751

Mental retardation and learning disabilities

► People with learning difficulties are at high risk for mental illness. Those with IQs of 50–70 account for 80% of people with learning disabilities. There is useful development of language, and learning difficulty only emerges as schooling gets under way. Most can lead an independent life. If IQ is 35–49, then most can talk and find their way about. In severe learning disability (IQ 20–34) limited social activity is possible. If less than this, simple speech may be unachievable. Special schooling and medical services are needed, as are adequate care and counselling for the families involved. In the UK, lack of resources and ambiguous community responsibilities are big major problems. medline 10548077 Further information: ask MENCAP (tel. ^{uk} 020 7454 0454).

*Antisocial personality disorder: www.erin.utoronto.ca/library/reserve/notes/FSC/FSC239Y/FORNSIC1.htm#dsm A key feature of psychopathy is deceiving and manipulating the unwary, while seeming sincere. Is this impression achieved solely via gestures and facial expression, or is their voice different? Acoustic analysis shows that psychopaths speak more quietly than controls with no tone changes related to emotionally-charged words [a9631561] [a1946892]

Managing psychopathy: beyond medicine and the law

Psychiatrists are sometimes unfairly criticized for abandoning psychopaths—as if they were too much trouble. This easy criticism does not take into account civil liberties: patients must either want treatment, or they must have a *treatable* mental illness before they can be detained against their will (or admission must be likely to prevent deterioration). Preventive detention is problematic as the rule 'innocent until proven guilty of a crime' is central to the requirements of justice.

Renewable sentences: It is against this background that the suggestion has arisen that those with a history of psychopathic violence should receive care outside current penal and health set-ups. Such people, it is argued by the National Association for the Care and Resettlement of Offenders (NACRO), could be detained at the end of their sentences if they are still thought to be a risk to others. For this to take place, courts would need to have new powers, and there is also a need for new specialist services. However, this is no guarantee against injustice occurring: for example, a man without psychopathy who poisoned his wife might be free to marry again after 10–15 years in prison, but a man with psychopathy who had held a hostage without harming them might never be free to try to rebuild his life.

Withdrawal of psychotropic drugs

Withdrawing benzodiazepines ▶ The withdrawal syndrome may well be worse than the condition for which the drug was originally prescribed. So try to avoid benzodiazepine use, eg relaxation techniques for anxiety, or, for insomnia, a dull book, sexual intercourse and avoiding night-time coffee may facilitate sleep. If not, limit hypnotics to alternate nights.

One-third of those taking benzodiazepines for 6 months experience withdrawal symptoms if treatment is stopped, and some will do so after only a few weeks of treatment. Symptoms appear sooner with rapidly eliminated benzodiazepines (eg lorazepam compared to diazepam or chlordiazepoxide). It is not possible to predict which patients will become dependent, but having a 'passive dependent' or neurotic personality appears to be partially predictive. Symptoms often start with acute anxiety or psychotic symptoms 1–2 weeks after withdrawal, followed by many months of gradually decreasing symptoms, such as insomnia, hyperactivity, panic attacks, agoraphobia, and depression. Irritability, rage, feelings of unreality and depersonalization are common, but hallucinations less so. Multiple sclerosis is sometimes misdiagnosed as these patients may report diplopia, paraesthesiae, fasciculation, and ataxia. Gut symptoms include D&V, abdominal pain, and dysphagia. There may also be palpitations, flushing, and hyperventilation symptoms. The problem is not so much how to stop benzodiazepine treatment, but how to avoid being manipulated into prescribing them unnecessarily. This issue is addressed on p 330.

Management of withdrawal • Augment the patient's will to give up (elaborate disadvantages of continuous prescribing). • Withdrawal is harder for short-acting benzodiazepines, so change to diazepam. • Agree on a contract that you will prescribe a weekly supply, but will not add to this if the patient uses up his supply early. • Withdraw slowly (eg by ~2mg/week of diazepam). Warn the patient to expect withdrawal symptoms, and not to be alarmed.

Withdrawing MAO inhibitors (eg phenelzine, tranylcypromine) Abrupt withdrawal of phenelzine leads to panic, shaking, sweats, and nausea in ~¼ of patients. Tranylcypromine has amphetamine-like properties and may be addictive.

Withdrawing tricyclics 4 withdrawal (discontinuation) syndromes (all rare)

- Cholinergic activation: abdominal cramps, D&V, dehydration.
- Insomnia, followed by excessively vivid dreaming on falling asleep.
- Extrapyramidal symptoms, eg restlessness and akathisia.
- Psychiatric symptoms: anxiety, psychosis, delirium, or mania.

Keep up treatment for ≥6 months post-recovery. Rate of withdrawal 25mg/5 days (amitriptyline). **NOTE** there are no reports of these withdrawal symptoms from non-tricyclic antidepressants (eg mianserin, trazodone).

Because of withdrawal symptoms it is tempting to stop treatment at the earliest opportunity—but this is a mistake. The severely depressed continue antidepressants for ~6 months after recovery. For amitriptyline, the maintenance dose during this phase of treatment is ~75mg/24h po. Further slow reductions should be made before stopping the drug.

Phenothiazines Withdrawal symptoms have not been reliably reported.

SSRIs Abrupt withdrawal of paroxetine (more than other SSRIs) can lead to deterioration in mood and cognition ± orofacial dystonias. More general SSRI withdrawal effects: aggression/impulsivity, nausea, lethargy, insomnia, headache. Very slow dose tapering may be needed to prevent symptoms. Another strategy is switching to fluoxetine which may suppress rather than cause discontinuation symptoms.

Introduction to the psychotherapies

As usual, it was dialogue that combed out my muddle. Arthur Miller *Timebends* 86

Medicine has three great branches: *prevention, curing by technical means and healing*—and psychotherapy is the embodiment of healing: a holistic approach in which systematic human dialogue becomes a humanizing enterprise for the relief of suffering and the advancement of self-esteem. Questions such as ‘what is the meaning of my life’ and ‘what is significant?’ are answered in a different way after exposure to a gifted psychotherapist. Changes occur in cognition, feelings, and behaviour. This is why psychotherapy is dangerous and exciting: it changes people. Hence the need for supervision and ongoing training and self-awareness on the part of the therapist.

Psychotherapy stands in stark contrast to the increasingly questioned technical, machine-based realm of medicine, and we accord it great prominence here, in the hope that our explicit descriptions, and their reverberations throughout our books will produce corresponding reverberations in our minds as we set about our daily work in *any* branch of medicine, to remind us that we are not machines delivering care according to automated formulae, but humans dealing with other humans. So, taken in this way, psychotherapy is the essence of psychiatry—and the essence of all psychotherapy is communication. The first step in communication is to open a channel. The vital rôle that listening plays has already been emphasized (p 320).

It is not possible to teach the skills required for psychotherapy in a book any more than it is possible to teach the art of painting in oils from a book. So what follows here (p 370–96) is a highly selective tour round the gallery of psychotherapy, in an attempt to show the range of skills needed, and to whet the reader’s appetite. It is not envisaged that the reader will try out the more complicated techniques without appropriate supervision.

The psychotherapies may be classified first in terms of *who is involved* in the treatment sessions: an individual, a couple, a family, or a whole group; and secondly they may be classified by their *content and methods* used: analytic, interpersonal, cognitive, behavioural. All psychotherapies are aimed at changing aspects of the patient.

Behavioural therapies (more details: p 370) aim to change behaviour, e.g. if avoiding crowded shops (agoraphobia) is the issue, a behavioural approach focuses on the avoidance-behaviour. Such approaches will define behavioural tasks that the patient is expected to carry out between sessions.

Cognitive therapy (p 370) focuses on thoughts and assumptions, promoting the idea that we respond to cognitive representations of events, not to raw events alone. If this is so, cognitive change may be required to produce emotional and behavioural change. So in the above example of agoraphobia the therapist would encourage articulation of thoughts associated with entering crowds. The patient might report that she becomes anxious that she might be about to faint—fearing that everyone will think her a fool. These thoughts would be looked at using a Socratic approach: ‘Have you in fact ever fainted? How likely would you be to faint? If someone fainted in front of you in a shop, what would you think? Are they foolish?’

Longer-term psychoanalytical therapies (p 378) are concerned with the origin and meaning of symptoms. They are based on the view that vulnerability to symptoms arises from earlier experiences and unresolved issues, e.g. from childhood. The therapist adopts a non-dominant stance encouraging the patient to talk without inhibitions. The therapist encourages change by suggesting interpretations for the content of the patient’s talk.

Which psychotherapy is most successful? This is tackled on p 386.

Definitions of psychotherapy

There are important differences in how people use the term psychotherapy. The first recorded definition states that

Psychotherapy includes every description of therapeutics that cures by means of the intervention of the psychical functions of the sufferer. (F van Eeden 1892 *Med Mag* i 233).

This definition is worth bearing in mind because, uniquely, it focuses on the content of the intervention made by the patient, not on the specifics of the therapist's intervention. The most general modern definition, and the one employed in this section, is summarized thus:

Psychotherapy denotes treatment of mental disorders and behavioural disturbances using such psychological techniques as support, suggestion, persuasion, re-education, reassurance, and insight in order to alter maladaptive patterns of coping, and to encourage personality growth. (Dorland's Medical Dictionary)

Some commentators feel that it is important to draw a distinction between counselling and psychotherapy—but using the above definition (or indeed any definition that recognizes the great heterogeneity of psychotherapy) no valid distinction can be made, unless it is between the various types of psychotherapy. The main issue to bear in mind is that psychotherapy can be categorized as more or less specific, and more or less involved in, and driven by, theory.

So is 'just being nice to patients' in the normal course of one's medical activities an example of psychotherapy at work? The answer is no—not because being nice is therapeutically neutral, but because one's attention is not focused on planning change through the systematic use of interpersonal techniques.

The issue of training is very important, and here are some questions that might usefully be addressed to anyone offering psychotherapy:

Is there proof of efficacy? Ask for evidence of long-term results.

What qualifications does the therapist hold? Is he or she supervised?

Is the recommended regimen tailored to the patient's unique needs?

How will progress be monitored?

Is confidentiality assured?

Is there support and follow-up after the formal programme ends?

Behavioural and cognitive therapies

Behavioural therapy is designed to treat symptoms such as phobias, obsessions, eating and sexual disorders, as well as more general anxiety and mild depression. In those with obsessive-compulsive disorders, rituals (e.g. hand-washing) respond better than ruminations. The therapy is not suitable on its own for those with schizophrenia, dementia, or severe depression.

The first step is to analyse in great detail the behaviour that the patient wants to change. If the patient has agoraphobia (say), find out what somatic and psychological symptoms he has, such as palpitations, and fear of fainting ('fear of fear'), and then relate these to exact points in the behaviour—e.g. on merely planning to go out, or on entering a particular shop. Next ask the patient to make a hierarchy of such situations, starting with the least threatening. Treatment starts with graded exposure to the least threatening situation (systematic desensitization) combined with teaching a method of anxiety reduction. This might be systematic relaxation, in which groups of muscles are stressed and relaxed in an orderly sequence. For example, while sitting in a chair, the toes are pressed firmly into the ground, and then the knees are pressed firmly together, and then the hands on the knees, and then the fingers of one hand into the opposite palm, each finger in turn. Each pressing lasts for about 30sec and is followed by a period of relaxation, and it is this which counteracts the feelings of anxiety as systematic desensitization through the hierarchy gets under way.

Concordance/compliance (p 460) needs fostering by providing written information about reasons for practising the new behaviours, and by explaining that major benefits of treatment only come after hard work. It is wise to enlist the help of the spouse to give encouragement at each achievement. Rituals such as hand-washing that is provoked by a particular stimulus (e.g. going to the toilet) are treated by repeated exposure to the stimulus while encouraging avoiding the ritual. The therapist illustrates what the patient has to do ('modelling'). Patients may be taught to interrupt obsessional thoughts, e.g. by doing a particular activity each time they start.

Cognitive therapy We respond to cognitive representations of events, not raw data. Mood and thoughts can form a vicious cycle. Using the example of depression: low mood leads to gloomy thoughts and memories (e.g. dwelling on exams you did badly at, rather than those in which you performed well). These gloomy thoughts make you feel more depressed (mood) and this lowering of mood makes your thoughts even more gloomy. Cognitive therapy tackles this circle by tackling the thoughts. Take, for example, the thought: 'I'm a failure, and all my friends are avoiding me'. In cognitive therapy the process is to: ●Clarify exactly what the thought is (do not let it be just a vague negative belief) ●Look for evidence for and against the proposition in the thought ●Look for other perspectives ●Come to a conclusion. 6 sessions are better than fewer. medline:112249

The therapist encourages the patient to find other explanations by challenging him, e.g. by examining what 'I'm a failure' really means. 'What are the important areas in your life?' 'What do you count as success or failure?' 'Catastrophizing' denotes the tendency to see disasters in small mishaps, e.g. 'he didn't like my hat; he doesn't like me; nobody likes me. There are many kinds of biased thinking that cognitive therapy helps patients to recognize, e.g. 'black and white thinking', over-generalizing (as in the case of regarding one failure as a symbol of everything).

Indications for treatment are many (all evidence-based): depression; neurosis; phobias; chronic fatigue; alcohol abuse; smoking cessation; insomnia; bulimia; chronic pain; psychosis, e.g. in challenging delusions. medline-96852

Evidence is increasingly available that CBT is as effective as pharmacological therapy for mild to moderate depression, with the added advantage that relapse is less likely. Is it possible to apply these lessons in primary care? The answer is Yes—but only if extensive training is offered. Randomized trials of bringing CBT to primary care with less extensive training (4 half-days) and with more realistic costs have been disappointing. In one study,* GPs' knowledge of depression, and attitudes towards its treatment showed no major difference between intervention and control groups at 6 months—ie training had no discernible impact on patients' outcomes. One reason for this may be that skills were indeed learned, but that there was insufficient time to apply these skills in ordinary general practice, where depression may only be one of a series of problems presented to the GP in a single 10 minute consultation. The lesson is: if the benefits of CBT are to be brought to primary care, what is needed is *time*: time to learn a new skill, and, above all, time to exercise it.

M King 2002 *BMJ* i 947; see also L Mynors-Wallis 1995 *BMJ* i 441

Group psychotherapy

The rationale underlying group psychotherapy is that the group provides an interactive microcosm in which the patient can be confronted by the effect of his behaviour and beliefs have on others, and be protected during his first attempts to change.* This implies that group psychotherapy (as with all psychotherapies) is only practical for those who want to change. It has also been found that the most suitable patients are: 1 Those who enter into the group voluntarily, and not as a result of pressure from relatives or therapists; 2 Those who have a high expectation from the group, and do not view it as inferior to individual therapy; 3 Those who have adequate verbal and conceptual skills. Patients who are unlikely to benefit include those with severe depression, acute schizophrenia, or extreme schizoid personality (the aloof, cold, hypersensitive introvert); hypochondriacs; narcissistic (very self-centred) or paranoid (suspicious and pessimistic about the rôle of others) patients; and sociopathic types (they have low thresholds for frustration and little sense of responsibility)—but sociopathy is notoriously difficult to treat by any means, and group therapy may be the least bad option, as other members of the group may provide the only valid mirror that can be held up to these people, and other people may provide the impetus for change.

Clearly the selection procedure needs to be carried out by an experienced psychotherapist. He will aim for a group of, say, 6–8 members balanced for sex, and avoiding mixing the extremes of age. He will decide if the group is to be 'closed', or whether it will accept new patients during its life. He will usually take on a co-therapist of the opposite sex, and he will prepare the patients in detail before the group starts. The life of the group (e.g. 18 months) will develop through a number of phases ('forming' → 'norming' → 'storming'). First there is a settling-in period when members seem to be on their best behaviour, seeking to be loved by the therapist, and looking to him for directive counselling (which he rarely provides). Next is the stage of conflict, as the patient strives to find his place in the group other than through dependency on the leader. Frustration, anger, and other negative feelings are helpful in allowing the patient to test the group's trustworthiness. It is worth learning that expressing negative feelings need not lead to rejection—and this is a vital prelude to the next stage of intimacy, in which the group starts working together.

Various models of group therapy are used, and typically the therapist will need to steer the group away from outside crises and searches for antecedent causes towards the here and now—eg by asking 'Who do you feel closest to in the group?' or 'Who in the group is most like you?' 'Who would you say is as passive (or aggressive) as you are?' But the therapist must avoid sacrificing spontaneity. He learns to use what the group gives him, e.g. 'You seem very angry that John stormed out just now'. He avoids asking unanswerable questions, especially those beginning with 'Why?'. His task is to encourage interaction between members and to facilitate learning and observation by members. Special methods used to augment this process include written summaries of group activities, video, and psychodrama.

Uses of group therapy Psychological conditions, eg addictions. medline:1079663

Breast cancer is an example. medline:1096433

Group therapy may be more available than other psychotherapies because it is cheaper and quite often just as good as, say, family therapy. medline:1074287

*Miriam was the threshing-floor on which he threshed out all his beliefs. While he trampled his ideas upon her soul, the truth came out for him... because of her, he gradually realized where he was wrong. And what he realized, she realized.' *Sons and Lovers*, JPS, 22.

Crisis intervention

Occupying the interval between the spilling of our lives and their congealing into history, crisis intervention recognizes that moments of maximum change are times of greatest therapeutic opportunity.* The heroic policeman who intervenes between the blade and the self's autotarget is, by this action, in the prime position, if he only knew it, to cut through to the heart of the matter, by entering into *dialogue* (and this takes courage too, for there are no rules of engagement, and your only tools are those your patient gives you). During the rescue you may concentrate on these questions:

- What events have led to the person's difficulties? Concentrate on his thoughts and actions in the last few days.
- What is his mental state at the moment (p 324)? It is vital to know about depression, suicidal ideas, and psychosis.
- When his mental state allows, get to grips with methods he has used in the past to combat stress and resolve crises.
- Who are the significant people in the person's life?
- What help can the person rely on from family and friends?
- What solutions has the person (or his or her family, school, or employer) tried in the present crisis? How have they failed?

Subsequent therapeutic strategies

- If the person has been very severely affected by the crisis, it will be appropriate to offer, and sometimes insist upon, his abandoning of his normal obligations and responsibilities. Temporarily relieving the patient of his responsibilities is necessary to allow concentrated contact ('intensive care') within a therapeutic environment—eg a hospital or crisis unit.
- Ensure by taking practical steps that the patient's commitments are adequately looked after (eg arrange transport of children to foster parents).
- Decide on the best way of lowering arousal (time spent talking is often preferable to administering anxiolytics, which may only serve to delay the natural process of adaptation). If the patient is shocked, stunned, or mute, take time to establish the normal channels of communication.
- As soon as the person is receptive, promote a sense of hope about the outcome of the crisis. If there is no hope (a mother, consumed by grief after losing all her children in a fire), then this too must be addressed.
- The next step is to encourage creative thinking about ways whereby the patient might solve the problems. Start by helping him think through the consequences of all options open to him. Then help compartmentalize his proposed solutions into small, easily executed items of behaviour.

As the immediate crisis passes, and the patient has reasonable psychological functioning, it will be necessary to put him back in charge of his own life. A period of counselling is likely to be appropriate. This is described on p 376. Making a contract about therapy is important in encouraging the patient to transfer from the 'sick rôle' to a self-dependent, adult rôle. Crisis intervention often focuses on loss of face, loss of identity, or loss of faith—in oneself, in one's religion, one's goals, or one's roots.

Meta-analyses suggest that crisis intervention is a viable part of home-care and can be used during the acute phase of any mental illness. medline:10796414

✱ *All home-care packages for severe mental illness need crisis management plans* Where implemented, this keeps the vulnerable in contact with staff (NNT≈13 over 1yr) and reduces family burden (NNT≈3), and is a more satisfying form of care for patients and families. It is also said to be cheaper.

*See Sylvia Plath 1963 *A Birthday Present* in *Ariel*, Penguin, page 50

Supportive psychotherapy

There are many people who need continuous psychotherapy, as they find daily activities pose unending stress. The smallest decisions are insurmountable problems, and the patient, lacking even a glimmer of insight, seeks support at every turn. What can we offer here?

- Reassurance—eg that his problems are not unique; he is not mad.
- Clarifying problems; instituting practical help (eg practising for job interview); explanation (eg about claiming benefits) and teaching how to recognize stress, and how to take the small steps to reduce anxiety (p 344)
- Positive feedback when he moves away from passive dependency.
- Encouragement in taking even the smallest step towards clearly defined goals—which should be set within his limitations.
- Help to change the patient's social world (job changes, protection from a hostile family, reducing social isolation).
- Counselling for relatives and friends. (Get permission first!)

Where tested against cognitive therapy, this less-sophisticated therapy sometimes comes out well, eg in longterm schizophrenia care, medline:10796390 and also in care of adolescents with major depression. medline:10632230

Counselling

Good novelists (and counsellors) are somehow large enough to embody the world—so their characters (clients) are not just recreated in their own image. *Nothing* human is alien to them. Such exercise of the imagination is what enables virgins to counsel prostitutes—which they *can*, if they are submerged in and are fully aware of human affairs outside themselves.

In its basic (non-directive) form this involves focusing on helping the person clarify what it is they want to do, and what would be needed to achieve it. It does not entail advice, which would run counter to the process of empowering the patient. Fundamental counselling skills are listening, understanding, and reflecting—(± summarizing, the use of silences, interpretation—p 378—and confrontation). Other points:

- History-taking: note how previous stress has been coped with.
- Production of an agreed full inventory of problems.
- Redefining problems in terms of attainable goals.
- Use of therapeutic contracts to negotiate small behaviour changes.
- Aim for adult relationships between patient, family, and therapist, eg with a contract *vis à vis* duties, frequency, and duration of therapy, and what is expected of the patient (*homework*), eg learning anxiety-reducing techniques, and carrying out *rewards*, eg cooking an extra-nice meal with the family if the patient achieves an anxiety-provoking task such as shopping.
- *Talking* out (not *acting* out) anger in safe but cathartic ways.
- Reassurance. The therapist must not only give overt reassurance, but also by his demeanour he must reassure the patient that *whatever* he reveals (eg incest or baby battering), he will not be condemned, but accepted. Note: this is one advantage of computer-based psychiatric history taking—the computer is not shocked when the patient tells of drinking two bottles of gin a day: it simply goes on to ask: 'What about whisky?'

Is being non-directive a way of evading responsibility? Is it dishonest to hide our attitudes? What is lost by being dispassionate? Compassion, perhaps? Note that not all counselling is non-directive. 'Problem-solving' models of counselling (OPPOSITE) are sometimes directive—and *may* be appropriate if the therapist knows the client well.

113 Counselling is a vital part of many packages of care (p 347). medline:9286179

Psychodynamic psychotherapy

Psychotherapy can be classified into three main types. **1) Embedded therapy** where therapy is part of a general package of mental health interventions, eg someone with depression might be prescribed antidepressants, be attending a Day Hospital, and receive individual psychotherapy. **2) Tailored therapy** An eclectic form of therapy tailored to the needs of the patient, often delivered by psychologists, eg a depressed patient offered some counselling sessions with a cognitive slant, followed by marital therapy. **3) Formal model-based therapy** The types of analytic therapy described below would be good examples. NB: not all patients benefit from brief, symptom-based cognitive therapy, and, equally, psychodynamic therapy does not help all conditions, eg schizophrenia. medline:10796437

Suitable candidates are those with ingrained maladaptive patterns of emotion and behaviour, eg personality disorder. medline:10518167 & 11136631

Here past experience, eg of intimate relationships with parents, influence the present functioning in unconscious ways. This basic idea derives from Freud and his followers such as Bowlby, Klein, and Winnicott.

Key concepts of psychodynamic psychotherapy include:

1) Psychological defences Just as the immune system protects our physical integrity, so psychological vulnerability is shielded by psychological defences. As with the immune system, overactive defences can lead to trouble. Primitive defences like repression (turning a blind eye to unpleasant experience) or splitting (dividing the world into black and white) can be maladaptive. We need to be able to think about and process experience if we are to learn from it. Thus a person who has been sexually abused as a child may as an adult enter abusive relationships without realizing the connection.

2) Transference and counter-transference The past patterns (transfers) our present reactions to people. If we have trusted our parents, we will be likely to trust our doctors, teachers, and friends. The intense psychotherapeutic relationship brings these assumptions to the fore where they can be examined, understood, and learned from. The doctor in turn has unconscious reactions to her patients based on the past, ie counter-transference. For example, if her mother was an alcoholic she may be oversolicitous, or rejecting, with alcoholic patients. The doctor's reactions are also a key to the patient's feelings: if a patient makes us feel rejected (as alcoholics often do), perhaps that person himself was rejected as a child and has turned to the bottle in compensation.

How does psychodynamic psychotherapy work? The therapist provides a secure *frame*—a regular time and place and her own consistency and acceptingness. The patient *narrates* vignettes about himself and his life—usually about three per session. The therapist *listens* carefully, both to the stories and to her reactions to them. She then makes *linking hypotheses*, or *interpretations* which offer *meaning* to the patient. Previously inexplicable behaviour begins to make sense. At the same time the patient forms a close relationship with the therapist based on *empathy*, *genuineness*, and *non-possessive warmth* (shown experimentally to be key therapeutic factors)—and, where necessary *challenge*. These may be novel experiences for the patient which can be *internalized*. The patient can *work through* his difficulties in the comparative safety of the therapeutic relationship, especially his reactions to *ending*, which will bring up previously unprocessed losses.

Psychodynamic therapy can be *brief* (brief dynamic psychotherapy)—suitable for circumscribed problems, eg unmourned grief, or *long-term*, which in the UK NHS is usually about 2yrs, but in private practice may be longer, and is suitable for patients with severe personality disorders.

We thank Dr J Holmes for this page; see Bateman & Holmes, *Introduction to Psychoanalysis*, Routledge

Sex therapy for couples

This is an example of *couple therapy*: often the problem is not specifically sexual, and sexual difficulties may recede once other aspects of the relationship improve. Here, specific sexual dysfunctions are considered in the light of a modernized *Masters & Johnson* approach using a model of sexual response entailing excitation, plateau, orgasm, and resolution. medline:9669216

Start with a full (joint) description of the problem. This may be premature (or delayed) ejaculation, female frigidity (anorgasmia), impotence, or dyspareunia (eg from spasm—vaginismus—or other physical causes). How did the problem start (eg after childbirth)? Was there ever a time when sex occurred as desired? Is the problem part of some wider problem? What does your partner expect from you? Are you self-conscious or anxious during sex? Are there medical problems, eg ischaemic heart disease, medline:10899274 or mastectomy; prosthesis use is discouraged at intercourse as it delays confrontation with and acceptance of mastectomy. Techniques of body imagery and sensate focus (below) have special rôles here. medline:10732534 & 1223313

Sexual history Early experiences; present practices; any hints pointing towards transexualism, commercial sex work, or drug abuse? Orientation to either or both sexes. Difficulties with other partners? When did you meet? What attracted you to each other?

Drugs Alcohol, hypotensives (impotence); *SSRIs* (delayed ejaculation); β -blockers, finasteride, the Pill, and phenothiazines (loss of libido). OHCM p 306: diabetes, cord pathology, prolactin \uparrow .

Principles of therapy and the approach to sexual difficulties comprise:

- 1 Defining the task which the couple wishes to accomplish.
- 2 Reducing the task to a number of small, attainable steps.
- 3 Asking the couple to practise each small step in turn.
- 4 At the next session, discussing difficulties encountered.
- 5 Ameliorating maladaptive attitudes.
- 6 Setting the next task.

(Both relate to performance anxiety, and vaginismus may be part of a *generalized* anxiety-defence mechanism medline:11716000). One sequence to agree with the couple might be:

- 1 A ban on attempted sexual intercourse (to remove fear of performance failure). Education and 'permission' giving (ie to talk about and engage in 'safe' sexual fantasies) is vital.
 - 2 Touching without genital contact, 'for your own pleasure', initially, with any non-genital part of the body, to explore the range of what is pleasurable, and then to concentrate on whatever erogenous zones are discovered ('sensate focus').
 - 3 Touching as above 'for your own and your partner's pleasure'.
 - 4 'Homework' using a vaginal dilator and lubricating jelly.
 - 5 Touching with genital contact, first in turn, later together. Problems in taking the initiative may now surface. In premature ejaculation, the partner stimulates the penis, and as orgasm approaches the man signals to his partner, who inhibits the reflex by squeezing his penis at the frenulum.
 - 6 Concentrate on playing down the distinction between foreplay and intercourse, so that anxiety at penetration is reduced.
 - 7 Vaginal containment in the female superior position so that she can stop or withdraw whenever she wants. She concentrates on the sensation of the vagina being filled.
 - 8 Periods of pelvic thrusting, eg with a 'stop/start' technique.
- : drugs (eg sertraline) may also improve ejaculatory control. medline:9598491

Sexual universals

There is a great deal more to helping people with sexual difficulties than is outlined opposite, which is just one approach to a common problem. Alternative psychodynamic approaches are equally valid.

In any therapy the following universals need to be addressed:

- Never assume that your patient is too old or too ill for sexual issues to be relevant. Assume that everyone has a sex life, albeit sometimes in fantasy only. Whenever it is explored, fantasy is always found to be an important component of sexuality. medline:7458662
- Psychological approaches are always important, whatever is offered by way of physical props or drugs such as sildenafil.
- Psychological events have physical sequelae, and physical events have psychological sequelae.
- All pleasure, including all erotic pleasure, is either purely sensory or arises from associations of ideas: this offers many points of intersection for negative operators, such as distraction, spectating, guilt, anxiety, fear of failure, pain, and inappropriate stimulation.
- All humans have a need to give as well as to take. Reawakening this instinct may be an important part of therapy. medline:9522407

Systemic practice (family therapy)

What used to be called *family therapy* is now better known as *systemic practice*, which is an evolving body of ideas and techniques focusing on a person's difficulties within the context of the people and culture that surround them. Therapy is based on the assumption that most people have the resources and potential for resolving life's difficulties. Therapists may work with individuals or families. Screening rooms may be used where co-therapists observe family interactions during therapy via a one-way screen.

Its origins began in *experiments*.¹ Behaviour maintains itself by feedback loops, eg disruptive behaviour in a son draws divorcing parents together. This led to *paradoxical intention*—the paradoxical approach where the symptom is prescribed so interrupting the behaviour–problem cycle. *Structural family therapy*,^{2,3} sought to be more objective. A family can be described in terms of dimensions. Research interviews have given rise to a measure of 'expressed emotion' (EE) which is associated with severity of chronic illness in many disorders (eg schizophrenia, anorexia nervosa, cystic fibrosis). Therapy includes prescribed exercises, eg parents may agree to go out for a meal at a secret location. In an over-involved family, this strengthens parental executive subsystems, providing opportunity for disengagement and management of concomitant anxiety. *Systemic family therapy* was pioneered by the Milan School, emphasizing family behaviour according to 'myths', 'scripts', and family secrets, which dominate the inter-generational transmission of repeating behaviours. Drawing a genogram (family tree) with the family is a good way to reveal these. Hypothesizing, maintaining a neutral stance and the use of circular questioning are important components.

*Narrative therapies*⁴ consider that knowledge is developed by storytelling rather than through the logico-scientific method, working on the basis that there is no single truth about the reasons for problems but that 'truths' are constructed via conversations between therapist and therapist. People can be maintained in problem-saturated lives by 'viewing themselves in the context of a dominant knowledge'. By constructing an alternative knowledge, they can be liberated to challenge the problem. Narrative therapists help by restoring people's control over their problems via externalizing the problem. Faecal soiling is popularly viewed as an act committed by the child as a response to family dysfunction. But if it is viewed as a struggle between the family and the problem by identifying the 'sneaky poo' as the enemy, then the family can be engaged in a battle against it. The use of written letters is also considered a useful tool.

*Problem solving therapy*⁵ makes use of a structured approach to draw on people's resilience, and motivate problem solving. It centres conversations on solutions, not problems. 'If it works, do more of it. If it doesn't work do something different. No problem happens all the time.'

Session 1

- 1 Why have you come?
- 2 How may we be of help?
- 3 The miracle question
- 4 Exception questions
- 5 Spectograms
- 6 Agreed achievable tasks

Session 2 and beyond

- 1 What's better?
- 2a Elicit: Ask about positive changes
- 2b Amplify: Ask for details on +ve changes
- 2c Reinforce: ensure client notices & values change
- 2d Start again. Ask what else is better.
- 3 Ending. How can you get back on track?

'If you woke up and a miracle had occurred in the night, how would you know? How would your life be different?'

Exception question: Search with the client for possible exceptions.

'On a scale of 0 to 10, how much would you like your miracle to happen?' 'What would have to happen/What would you have to do to make your score move from 3 to 4?'

Family structure and functioning

Family structure is viewed by Minuchin³ as an invisible set of functional demands that organize family interactions. These transactional patterns are self-regulating in a way that attempts to return a family to its habitual mode and minimize anxiety.

Dimensions of family functioning⁶ (McMaster model) The 6 dimensions allowing any practitioner to describe family functioning are:

- **Problem solving:** Can the family act together to solve everyday emotional and practical problems? Can they identify a problem, develop, agree, and enact solutions and evaluate their performance. Success dependent upon functioning in other dimensions.
- **Congruence of verbal and non-verbal communications:** Are communications clear and direct or are there hidden agendas or hidden meanings? Do people listen to one another?
- **Roles:** Who is in charge and how are executive decisions made? Who provides for the family? Who is concerned for the child's education and emotional development? Families may function most effectively when rôles are appropriately allocated and responsibilities explicit.
- **Affective involvement:** Relationships in families tend to exist on a continuum between over-involved (enmeshed) to disinvolved (disengaged). Empathic involvement is ideal. This depends on development, as greater involvement is needed for babies than adolescents. Enmeshment may lead a child to be so anxious about a parent that they feel unable to leave them, and avoid school as a consequence.
- **Affective responsiveness:** How do individual family members respond emotionally to one another both by degree and quality? Welfare feelings would include love, tenderness, and sympathy. Emergency feelings would include fear, anger, and disappointment.
- **Behavioural control:** How is discipline maintained? Is there negotiation? Is it flexible? Chaotic? Absent? (depends on quality of communication).

Dysfunctional family patterns: triangulation. When parents are in conflict, each demands the child sides with them. When the child sides with one, they are automatically considered to be as attacking the other. The child is paralysed in a no-win situation where every movement is defined by one parent as an attack.

Scapegoating When an individual is singled out by the family as the sole cause of the family troubles. This serves to temporarily bury conflicts that the family fear will overwhelm them.

↑ Expressed emotion: Derived from a family interview: reflects hostility, emotional over-involvement, critical comments, and contact time.

We thank Justin Williams and Bill Ness for providing this page.

See also: Efficacy of brief dynamic psychotherapy for children with emotional disorders 2002 *Psychother Psychosom* 2002 71 28 [11740166]

1 G Barnes *Family Therapy* 946, in M Rutter 1994 *Child and Adolescent Psychiatry* 3rd, Blackwell

2 D Will 1985 *Integrated Family Therapy*, Tavistock, London

3 S Minuchin 1974 *Families and Family Therapy*, Tavistock, London

4 M White 1990 *Narrative Means to Therapeutic Ends*, Norton, New York

5 S De Shazer 1985 *Keys to Solution in Brief Therapy*, Norton, New York

6 N Epstein 1981 *Problem-centred Systems Therapy* in A Gurnian *Handbook of Family Therapy*, New York, Brunner/Mazel. See <http://solutionmind.com/solutiongroup/rapy.htm>

and H Klar 1995 *Pediatr Clin North Am* 42 31 Medline:[7854868]

Play therapy*

The people who have most experience in psychotherapy with children are parents. They hold the key cards for influencing a child's behaviour. These comprise love, mutually understood channels of communication, systems of valid rewards, and a shared knowledge of right and wrong. It is the families without these which are most likely to need the help of professionals. The following guidelines are offered to help these children:

▶ Never underestimate a child's capacity for insight: if there is lack of insight, it may be your own: *do not expect children's methods of communicating insight to mesh with adult's*. Play, rather than talk, may be its medium. medline:11144854

- Take time early on to make friends with the child. Don't rush.
- Accept the child on his own terms—exactly as he is.
- Avoid questioning, praising, or blaming. Be totally permissive.
- Don't say 'Don't', and only restrain if about to harm himself.
- Show the child that he is free to express any feeling openly.
- The responsibility to make choices is the child's alone.
- Follow where the child leads: avoid directing the conversation.
- Use whatever he gives you. Reflect his feelings back to him.
- Encourage the child to move from acting-out his feelings in the real world, to expressing them in words and play.
- Prepare the parents for change in the child.

In play therapy the child and the therapist play together with toys which give the child an opportunity to verbalize his innermost fantasies. As Virginia Axline explained to one of her 5-year-olds,² play therapy is 'a time when you can be the way you want to be. A time you can use any way you want to use it. A time when you can be you.'

Verbatim example of play therapy in action, with 6-year-old Dibs

He walked around the playroom. Then he picked up a doll. 'Well, here is sister. I'll get her to eat some nice rice pudding, only I'll poison her, and she'll go away. . . forever and ever.' 'You want to get rid of the sister?' the therapist remarks. 'Sometimes she screams and scratches, and hurts me and I'm afraid of her. . . She's five now . . . She'll go to the same school as I go to next year.' 'And how do you feel about that?' 'Well, I don't care. . . she doesn't bother me like she used to.' He went over to the easel and picked up a jar of paint, adding further colours and stirring it well. 'This is poison for the sister. She'll think it is cereal and she'll eat it and then that will be the end of her.' 'So that is poison for the sister and after she eats it, then that will be the end of her?' Dibs nodded 'I won't give it to her yet a while, I'll wait and think it over.'

Before play therapy Dibs had been sullen, mute, and unco-operative for years on end. No one had ever been able to engage him in normal family or school activities—an impossible child—until play therapy, by allowing him to express negative feelings, unlocked those reserves of resilience, creativity, and zest for life that lie dormant or otherwise, in every child.

play therapy is not the sum of child psychotherapy: the above is given simply to illustrate the issues involved. Also, play is a prominent part of therapies that are not play therapy: play can prepare children for procedures such as micturating cystograms and MRI examinations. medline:9413970

Play is also a valuable way of promoting self-care in diabetes. medline:9128656

Play therapy has also been validated as a component in treatment of obsessive-compulsive disorder and childhood sexual abuse. medline:10553460 & 11185951

*V Axline *Play Therapy*, Ballantine and Dibs: *in Search of Self*: [[@10616294](#)]

Art or art therapy?

'The painting group provides quick access to the emotions and it permits the establishment of a strong therapeutic relationship.'

medline:11234248

For many people, art simply soothes and placates a tortured, tired, or timid mind. But art may go further even than healing (p 465), by approaching humanity by way of self-knowledge—a route that may entail wounding in equal measure to repair. Researchers have shown, seemingly idiotically, that reading poetry relieves stress. This is rather like finding out that if plants are put out in the rain they get wet—whereas the real conclusion is that rain makes them grow. It is this propensity to generate growth and development which is the essence of art, and, as we have observed elsewhere, growth and development are the essence of childhood. This implies that there is some relation between art and our pre-verbal, pre-logical life. We all once knew how to paint and draw: a well of knowledge that has, for most of us, been pierced by the reductionist logic of our mature years, so that this ability, like life itself, has leaked away, without our really knowing how or why or where. So we look into the well and it's empty.* The answer is to dig deeper, not to try to move the well to a better site—for as the ancient proverb states: *you can move a city, but you cannot move a well.*

Are these fascinating or futile observations of any use to our patients? To answer this question we might do best to look to two extremes: those individuals with a logico-verbal deficit (eg those with some forms of schizophrenia), and those with logico-verbal excess (eg overworked students). In the first category, art therapy has a much talked of relevance. Vittorino Andreoli has pointed out that artistic expression is a form of non-verbal communication that, once decoded, is most relevant to therapy and communication with the therapist. 'In pathologies where there is pronounced logico-verbal deficit, such as schizophrenia, this form of non-verbal communication becomes essential.' (P Pini). Notice that this art arises out of the need to speak, not the need to represent. www.rawvision.com/back/carlo/carlo.html

As for the rôle of art in those with logico-verbal excess, suffice it to recall the words of the greatest logico-verbal exponent of all time, Marcel Proust, who tells us, in *La Prisonnière* that '... Music, very different in this respect from Albertine's society, helped me to descend into myself, to make there a fresh discovery. ... As the spectrum makes visible to us the composition of light, so the harmony of a Wagner, the colour of an Elstir enable us to know that essential quality of another person's sensations into which love for another person does not allow us to penetrate. ...'

*WH Auden (1937)—*As I walked out one morning:*

*In headaches and in worry
Vaguely life leaks away,
And Time will have his fancy
To-morrow or to-day ...*

*It was late, late in the evening
The lovers they were gone;
The clocks had ceased their chiming,
And the deep river ran on.*

From 'Tell me the truth about love'

Comparing the psychotherapies

► Is it true that 'Everyone has won and all must have prizes'? medline:239666

One reason why it is hard to demonstrate that one form of psychotherapy is better than another is the high proportion of patients (~80%) who benefit from almost any intervention. A small (but perhaps useful) difference between two treatments is likely to be swamped by the large proportion of patients who will benefit from either technique.

The methodological problems of comparing different studies of varying validity have now been addressed in a systematic way (using techniques that themselves are not beyond criticism) and yield the following 'conclusions' from research that has met most of the ideal design criteria: randomness; independent assessment; therapy being given by comparable, experienced therapists working for equal amounts of time on their respective patients; well-matched groups; and adequate sample size.

Which is better? medline:239666

	First item	Second item	Tie
Psychotherapy vs minimal or no treatment	9	0	5
Individual vs group psychotherapy	2	2	9
Time limited vs time unlimited psychotherapy	2	1	5
Non-directive vs other psychotherapies	0	1	4
Behaviour therapy vs analytic psychotherapy	6	0	13
Drugs vs psychotherapy	7	0	1
Drugs vs psychotherapy and drugs	0	6	5
Medical régime alone (eg for asthma) vs psychotherapy plus medical régime	1	9	1

The chief finding is a large number of ties, except when psychotherapy is compared with minimal treatment (it is better) and when it is combined with drugs, which confer a more-or-less consistent benefit. medline:10796818

The diagnoses in the above table were mixed (ie depressed, neurotic, or schizophrenic, with rather over-inclusive USA criteria), and so the question arises as to whether patients with different diagnoses respond differently to psychotherapy. The few consistent findings are that behavioural-cognitive therapy is well suited for treating circumscribed phobias, obsessive compulsive, sexual, and habit disorders, and that cognitive therapy is good for depression and may be better than antidepressants in preventing relapse; this is also true for socially phobic alcoholics. medline:10890288 &: 10773778

It may also be more helpful than behaviour therapy in bulimia. Family intervention is known to be particularly helpful in preventing relapse in emotional families with a schizophrenic member.

It is hard to quantify improvement with psychodynamic therapy (psychoanalysis and its descendants), but there are convincing trials of its use in helping with chronic disease, eg irritable bowel syndrome medline:8401959 but in other cases, therapy may last for years, so outcome studies are expensive and difficult. So purchasers of health care (p 435) are reluctant to fund this type of care; but this should not blind purchasers to the real and large benefits randomized trials have 'proved' in the well-focused, usually brief psychotherapies above. medline:7950744

Index on child mental health problems

The next section (up to p 396) deals with some aspects of childhood mental health—but many issues in child psychiatry overlap with pages in the adult section of this chapter, and also with paediatric practice. The psychiatry of attempted suicide is a good example. Many of these patients will be in the last phases of childhood, and it is unclear which service will suit them best. As ever, take a holistic view of your patient, and design a care plan which takes these facets into account.

- Abuse, p 284
- Alcohol abuse, p 363
- Anorexia and bulimia, p 348
- Attention deficit disorder, p 207
- Asperger's syndrome, p 742
- Autism, p 392
- Bed wetting, p 206
- Bullying, p 394
- Cultural issues in mental health symptoms, p 436
- Drug addiction, p 362
- Dyslexia, p 396
- Encopresis, p 206
- Enuresis, p 206
- Family therapy, p 382
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- Psychosis and psychosis-like symptoms in children, p 390
- School refusal, p 207
- Sleep disorders, p 390
- Substance abuse, p 362
- Suicide, p 338
- Talking to young people about sex, p 328
- Tics and Gilles de la Tourette syndrome, *OHCM* p 726

Depression in children and adolescents

Prevalence Depression affects at least 2% of children under 12 years, and 5% of teenagers, much going unrecognized.

Presentation is often with somatic symptoms \pm anxiety.¹ Sometimes presentation is simply as poor functioning at school, socially, or at home, even masquerading as bad behaviour (particularly in boys). Mood is characteristically much more variable and less pervasive than adults—rapid swings commonly occur. The fact that children are able to enjoy some aspects of their life shouldn't preclude the diagnosis of depression.

Features as seen in adults:

Low mood
Loss of interest
Socially withdrawn
Poor self-esteem
Psychomotor retardation
Tearful
Guilt
Anxiety
Lack of enjoyment in anything

Features common in childhood:

Running away from home
Separation anxiety \pm school refusal
Complaints of boredom
Poor school performance
Antisocial behaviour
Insomnia (often initial, not early waking)
Hypersomnia
Eating problems

Young children may present with sadness and helplessness; slightly older children with feelings of being unloved and unfairly treated; whilst guilt and despair may be more prominent in teenagers.*

Interviewing is usually best conducted one-to-one. Consider the possibility of concealed factors (eg past child abuse, bullying). Parents can be interviewed separately (ask the child's permission, and see the child alone, again, if appropriate, *after* seeing the parents, to report back: this helps avoid the impression that you are somehow colluding with the parents). NB: parents may not always be aware of depression in their children. Assessment is often difficult: questions may only be answered by silence or a shrug. If not getting anywhere, talk to the child—or just offer silences—but do not give up.

Always ask about suicidal ideas and thoughts of selfharm. Note any past attempts: ~15–20% make further attempts (excess risk if: male, conduct disorder, \uparrow alcohol use, hopelessness, and those in local authority care). Self harm can be regarded as a form of communication—a message in a bottle; not always 'picked up' and sometimes it is difficult to decipher the teenager's exact intentions.* Refer urgently if risk is considered significant. Adolescents with conduct disorders can be manipulative and extremely difficult to assess—an urgent second opinion is frequently of help.

Management—ideally a combined approach:

- Social interventions—addressing any sources of distress (eg bullying) and removing opportunities for self-harm (eg paracetamol at home).
- Psychological interventions—encourage the patient to talk about their fears and anxieties. Counsellors, sympathetic teachers, and youth workers may be a suitable resources. Cognitive behavioural therapy has been shown to be of help—but may not be freely available in primary care.
- Drugs: often disappointing (and dangerous); *fluoxetine* may be the least bad. Beware of behavioural problems. *Tricyclics* are less good. medline:0908557 & 11806021 Where there is a marked sleep disturbance, mirtazapine may be less stimulating, and effects on sleep start immediately.*

* Q Spender et al. *Child Mental Health in Primary Care*, Radcliffe Medical 2001 ISBN 1-85775-262-7
See also SJ Kingsbury; *Parasuicide in adolescence a message in a bottle* ACP Rev Newsletter 15:253–9

Psychosis-like symptoms in childhood

In many centres, early-onset schizophrenia is diagnosed with the same criteria as adults (p 358) as it appears to be continuous with later onset forms (with a preponderance of males being affected). medline:11434484 This may be over-simplistic, as hearing voices and delusional beliefs are not uncommon in childhood and have a rôle in augmenting the worlds of play and make-believe. They may also be culturally mediated, eg in some Jewish communities where demons are personified. medline:11383445 Another example is delusional erotomania (p 744) which is reported to be common in adolescents living in places such as China where expressing sexual interest has to be indirect. There are many other examples where to equate delusions and hallucinations with psychopathology would be wrong. medline:8894203

If hallucinations *do* reflect psychopathology, the diagnosis may be obsessions rather than psychosis, as in the unwanted hearing of tunes.

Hallucinations are also more common in the isolated and withdrawn; here their importance may lie in alerting you to this fact. medline:9895209 & 10803816 Some hallucinations should receive very serious attention:

- Those which are imperative ('kill so-and-so')
- Those heard as if outside the head
- Those exciting strong emotions
- Those referring to ideas that the person feels are not their own.
- Multiple voices talking at once, and especially voices talking to *each other*.

Sometimes hallucinations resist diagnosis. This is not in itself a problem as the diagnosis will sooner or later become clear. Meanwhile, ask yourself whether these odd ideas are likely to indicate that your patient is at increased risk of serious outcome, eg suicide. medline:10714054 & 11417259

Causes of odd ideas Substance abuse; drugs; schizophrenia; anxiety/depression; hypomania; head injury; epileptic aura; migraine; Charles Bonnet syndrome (p 500); SLE; encephalopathy (eg lead exposure); infections (herpes, EBV-associated Alice-In-Wonderland syndrome); stress; physical abuse.

Tests MRI/CT may be indicated, eg in olfactory hallucination. medline:11167318

Management ▶ Early intervention helps, and may reduce chances of later chronic schizophrenia, so refer promptly. medline:11167318

If you are the child's GP, ensure there is a treatment plan with a named worker, incorporating antipsychotics (if indicated, p 360) with psycho-educational, psychotherapeutic, and social components. New antipsychotics (p 361) are rarely *specifically* licensed for children, but their use in well-monitored environments is encouraging. medline:11642479 are legion; they may not be as bad as older drugs. medline:11603883 & 11461715

Prognosis Spontaneous improvement of psychotic-like symptoms occurs in the majority of children. In one follow-up study, many developed chronic mood disorders; <50% met diagnostic criteria for a major disorder (schizoaffective or bipolar disorder, depressive disorder, 'psychotic disorder not otherwise specified'). In those not developing a mood or psychotic disorder, disruptive behaviour disorders are very common. medline:11458307

Sleep problems and the parasomnias

Waking at 3am—ready to play, or wanting entry to the parent's bed. For those not appreciating these visitations from the pure of heart, the easiest thing is to refuse to play and buy ear plugs to lessen the impact of screaming—or to let the child into the bed. Or try extinguishing the behaviour by attending the child ever more distantly: cuddle in bed → cuddle on bed → sitting on child's bed → voice from doorway → distant voice. Try to avoid hypnotics. If drugs are essential, consider triclofos sodium 25–30mg/kg.

Other sleep disturbances. Hunger/colic (infants); poor routines (preschool); worry (adolescence). Bedroom tv may be to blame. www.txt.co.uk/full.cfm?1039 Try behavioural therapy before hypnotics. **Causes:** night sleep ↓; depression; sleep apnoea (*OHCM* p 190); narcolepsy (*OHCM* p 724); and **hypersomnia** (rare in children): suspect this whenever sleepiness occurs with extrapyramidal effects, oculogyric crises, myoclonus, inversion of diurnal rhythms, obsessions, and mood change. The cause is said to be viral (influenza, rarely measles), but Q fever and mycoplasma have also caused this syndrome, as has infiltration of the hypothalamus (eg by lymphoma). MRI: look for subcortical involvement.

Sleepwalking and parasomnias Of all our non-insane automatisms, somnambulism is the most familiar and striking, literally, sometimes, as households may occasionally be endangered when the bloodiest dreams of junior somnambulists are enacted. The young are by far the most proficient sleepwalkers (the old may emulate them if stressed, esp. if this is associated with excess alcohol or caffeine use, and lack of stage IV sleep—our deepest sleep). Any psychic event associated with sleep may be termed a parasomnia.

Parasomnias comprise: ● **Arousal disorders** (sleep walking, night terrors, and 'confusional arousal') ● **Rhythmic movement disorders** (rhythmic movement head-banging disorder) ● **REM sleep disorders** (rapid eye movement sleep associated nightmares, sleep paralysis, hallucinations, and REM sleep behaviour disorders, involving motor enactment of dreams, sometimes with injury to oneself or others) ● **Miscellaneous disorders** (sleep-talking; nocturnal enuresis; myoclonus nocturnus; and nocturnal bruxism, ie teeth grinding).

Suffering from night terrors is often a familial problem. The child awakens frightened, hallucinated, and inaccessible—and is obviously alarmed.

It is common to observe movement in children during sleep: it is their repetitive nature which allows the diagnosis of rhythmic movement disorder. The movement may be body rocking, leg rolling, or head-banging (this 'jactatio capitis' may lead to subdurals, fractures, eye injuries, and false accusations of abuse). Tongue-biting may suggest epilepsy. But do not try to be too obsessive in differentiating parasomnias from nocturnal epilepsy, for three reasons: the first is that our definition of epilepsy is tested to destruction by the parasomnias ('epilepsy is intermittent abnormal brain activity manifesting as simple or complex seizures'). The second reason is that those with clearly defined parasomnias are at risk of developing tonic-clonic nocturnal seizures later in life. The third reason is that some parasomnias are signs of autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE). ADNFLE is associated with abnormalities in the gene encoding nicotinic acetylcholine receptor α_4 -subunit (chromosome 20). In these individuals, correct diagnosis may be aided by EEG (rhythmic slow anterior activity) and video-polysomnography (shows sleep-related violent behaviour, sudden awakenings with dyskinetic or dystonic movements, and complex behaviours ± enuresis). medline:9549500

Bedtime clonazepam; amitriptyline; carbamazepine. medline:7969657
If these do not work, self-hypnosis may help, or the simple expedient of waking the patient 30min before the expected parasomnia.

Sleep paralysis

When we sleep we trawl forbidden seas, arranging and being rearranged by the flotsam and jetsam of our waking lives. As our nets descend through the various stages of sleep, our Sovereign Reason or Will usually remains quietly on deck, but if by chance it descends with the nets then, like the tail wagging the dog, it adopts and propagates a life of its own, which may be full of danger. Because our vessel has been vacated, Marie-Celeste-style, we are judged, in Law, not to be responsible for our actions while asleep—be they theft, arson, or homicide. Without will or wind, our vessel, like Coleridge's *Mariner's*, is moved 'onward from beneath' by secret forces.* So here we have the model of Reason and Will residing, during sleep, either upstairs or downstairs: but in sleep paralysis, neither is the case—the tail cannot wag the dog, nor the dog wag his tail. Sleep paralysis may involve complete paralysis of all voluntary muscles, even the diaphragm. For anyone who has experienced it, it is frightening, unforgettable, and difficult to describe—like being aware during anaesthesia. Sleep paralysis was first described by Ishmael during a reverie on deck, sleeping between Queequeg, the cannibal whaler from the South Seas, and his harpoon, before Herman Melville embarks them on the *Pequod's* voyage to track down Moby Dick '... At last I must have fallen into a troubled nightmare of a doze; and slowly waking from it—half steeped in dreams—I opened my eyes, and the before sunlit room was now wrapped in outer darkness. Instantly I felt a shock running through all my frame; nothing was to be seen, and nothing was to be heard; but a supernatural hand seemed placed in mine. My arm hung over the counterpane... for what seemed like ages piled on ages, I lay there frozen with the most awful fears ... thinking that if I could but stir it one single inch, the horrid spell would be broken...'

* *Till noon we quietly sailed on,
Yet never a breeze did breathe:
Slowly and smoothly went the ship,
Moved onward from beneath.
Under the keel nine fathom deep,
From the land of mist and snow,
The spirit slid: and it was he
That made the ship to go.*

Samuel Taylor Coleridge Part v of
the Rime of the Ancient Mariner

Autism

This neurodevelopmental disorder is, if severe, an antithesis of our definition of mental health (p 314). *Prevalence*: 1:500 of those <16yrs; estimates vary. ♂:♀≈4:1. *Genetics*: complex. *medline*:10625077 Autism is a triad of:

- 1 Impaired reciprocal social interaction (A symptoms).
- 2 Impaired imagination, associated with abnormal verbal and non-verbal communication (B symptoms).
- 3 Restricted repertoires of activities & interests (C symptoms). *medline*:9519632

Diagnosis: Try to identify >8 of the following symptoms—and these should include at least 2 'A' symptoms; 1 'B' symptom and 1 'C' symptom. (Telling comments before the age of about 2 are: he does not respond to his own name; he hates his routine being changed; he is not interested in toys.)

Impaired reciprocal social interaction (A symptoms)

- Unawareness of the existence and feelings of others (treating a person as a piece of furniture; being oblivious to others' distress or need for privacy).
- Abnormal response to being hurt (he does not come for comfort; or makes a stereotyped response, eg just saying 'Cheese, cheese, cheese').
- Impaired imitation (eg does not wave 'bye-bye').
- Abnormal play: eg solitary, or using others as mechanical aids.
- Bad at making peer friendships. If he tries at all, the effort will lack the social conventions, eg reading the phone directory to uninterested peers.

Impaired imagination (B symptoms) associated with abnormal communication

- No babbling, facial expressions or gestures in infancy.
- Avoids mutual gaze; no smiles when making a social approach; does not greet his parents; stiffens when held.
- Does not act adult rôles; no interest in stories; no fantasy.
- Odd speech production (eg echolalia, ie repetitions); odd use of words ('Go on green riding' for 'I want a go on the swing'); pronoun misuse ('You' instead of 'I'); irrelevances (odd interjections).
- Difficulty in initiating or sustaining conversations.

Poor range of activities and interests (C symptoms)

- Stereotyped movements (hand-flicking, spinning, head-banging).
- Preoccupation with parts of objects (sniffing or repetitive feeling of a textured object, spinning wheels of toys) or unusual attachments (eg to coal).
- Marked distress over changes in trivia (eg a vase's place).
- Insists on following routines in precise detail.
- Markedly narrow range of interests, eg preoccupied with lining up objects; or in amassing facts about the weather.

For the *CHAT* test, see: www.feet.org/autism/chat-test.htm. *CHAT* has a sensitivity of 38% and a specificity of 98% if done at 18 months old. *medline*:10846303

Mostly unknown; ?prenatal rubella; ?tuberous sclerosis. Concordance in monozygotic twins: 90% (twin studies are bias-prone *medline*:11780600).

There is no cure. Behaviour therapy may be tried. ▶A good teacher is more helpful than a good doctor. Special schooling may not be needed (but IQ may be ↓). Learning may be better by overhearing than by direct methods. Joining a self-help group is wise. UK phone: 020 81830 0999. Encourage parents to give more attention for 'good' behaviour rather than 'bad', and to have rules for behaviour. 70% remain severely handicapped. 50% develop useful speech; 30% develop seizures by adulthood. 15% will lead an independent life. Apply for benefits (disability living allowance in UK).

Parents will ask about secretin; trials don't support its use. *medline*:10588965

Bullying

▶ Every individual should have the right to be spared oppression and repeated, intentional humiliation, in school as in society at large.* Bullying is important not just because it is unacceptable to the child concerned, but also because it leads to depression, somatization, drug abuse, and suicide. medline:11182867 Bullying is also bad for bullies: antisocial behaviour persists into adulthood with impaired reciprocal diadic relationships (≈poor love life). medline:11232258

Incidence 27% of primary and middle school children report bullying each term, and 10% in secondary pupils. In a study of prevalence, 4% were direct bullies, 10% bully/victims (both bully and victim), and 40% victims. Pure victims, especially girls, were most likely to have physical health symptoms (eg repeated sore throats, colds, and coughs). Pure bullies (who never got victimized) have the least physical or psychosomatic health problems.**

Risk factors Isolation; being small; being gay or seeming gay. medline:10698879

Assessment Whenever a child is stressed, depressed, or suicidal, suspect bullying. **Signs and symptoms** Hyperventilation, vomiting, accidents, back pain, abdominal pain, bruising, submissive behaviour, school refusal.

Management Interventions that target either bullying or emotional distress may reduce the severity of both problems. medline:11260830

Liaise with the school. Psychotherapy may have a rôle. Action needs to be taken to ensure that the bully doesn't prosper from bullying, so learning to 'achieve dominance over others by the abuse of power'. medline:7866165

The hardest task is to combat the ethos among bystanders, which allows bullying to continue as if it were 'none of my business'. medline:7787625

This implies that we all have a rôle in minimizing bullying. Bullying in the health services is well-documented,** and we have probably all been affected by frightened people abusing their positions of power. We fear to act when we are bullied because we might get a bad reference or because we don't think our own humiliation is that important. It is easier to take a more rational view when we see others being bullied—we simply must not allow it, and recognize that the bullier may not be able to stop the behaviour without help. Help is available: see www.nhs-exposed.com

Prevention School-based anti-bullying intervention programmes are successful in primary schools but less so in secondary schools. medline:10900778

Vis-à-vis homosexual bullying, research suggests that teachers are aware of homophobic bullying but are confused, unable, or unwilling to address needs of the pupils concerned. Perhaps citizenship education programmes have an important rôle here. medline:11259075

Problems may become very deep-rooted. Studies of bullying by teachers reveal the subtlety and complexity of teachers' strategies for distancing themselves from being held accountable for intimidation. medline:11190687

On a more universal plain, perhaps the first step is to recognize that we ourselves are potential bullies, and these traits come to the fore when we are stressed, frightened, overworked, and threatened by uncontrollable events. We stop ourselves from being bullies, more or less successfully, by intrapsychic appeals to our well-respected mentors who 'would never behave like that', and by communicating our feeling to our colleagues directly before they are forced underground only to resurface as bullying. Try: 'I'm feeling rather stressed at the moment: tell me if I seem bullying or hectoring—but we've got to get this job done, and I suggest doing it like this. . .'

* L Quine 2002 *BMJ* i 878 & D Olweus *Bullying at school*. Oxford: Blackwell, 1993

D Wolke 2001 Bullying in primary school and health problems *Arch Dis Chi* **85 197

****BMJ* 2001;323:1314 (1 Dec) www.bmj.com/cgi/content/full/323/7324/1314

Dyslexia

Reading ability usually goes hand-in-hand with intelligence, but when this is not the case, and someone with, say, an IQ of 130, finds reading difficult, the term dyslexia is often used. The term 'specific learning difficulty' is preferred by some people, as 'dyslexia' is often a term used by parents to help them cope with having a child whose general intellectual skills, including reading, are less than hoped for. NB: dyslexia can be associated with other speech and language disorders: consider referral to a speech and language therapist (SALT).

There is a problem with appreciating phonemes, eg that 'cat' comprises /c/, /a/, and /t/. Breaking up unfamiliar words into phonemes and having a go at stringing them together is the central act of learning to read. This is what needs to be taught, educationalists say, and children should not be made to rely on unstructured guesswork (the 'look and say' approach) which is now discredited as the sole means of equipping children for reading.

Quite often, distortion/jumbling of text during reading is reported. Visual aids have been used with some success to improve reading. medline:10561921

Children with dyslexia also have difficulty in telling how many syllables there are in a word (don't we all?—'How many syllables are there in *strength*?' analytical dyslexics are prone to ask). They also have difficulty with verbal short-term memory—eg for meaningless strings such as telephone numbers. There is also a problem with telling if two words rhyme or not, and in distinguishing phonemes which sound similar (eg /k/ and /g/). There may also be left/right muddle.

Boys are more afflicted than girls, and show stronger genetic effects (up to 50% of boys are dyslexic if their fathers are). Genes on chromosomes 1, 2, and 15 are implicated, and linkage on chromosome 6 near the human leukocyte complex may explain associations between dyslexia and autoimmune diseases. medline:7939663

Rarely, CNS examination reveals a left-sided cerebral lesion. Usually changes are only seen at *postmortem*, eg in perisylvian regions \pm unusual asymmetry of the plenum temporale (Wernicke's area), with cortical dysplasia and scarring. *In vivo* characterization of this asymmetry is becoming possible. medline:10443884

Perhaps dyslexia reflects weak connectivity between anterior and posterior parts of the language areas of the brain, and the angular gyrus. Positron emission tomography shows that when dyslexic adults perform rhyme judgments and verbal short-term memory tasks they activate less than the full set of centres normally involved with these tasks. www.shianet.org/~reeneenew/hist.html

Going along-side these theories is the functional MRI observation of specific involvement of one subsystem of the visual pathways which prevents rapid processing of brief stimuli presented in quick temporal succession. This is the so-called medline:11099442

Make sure the 'dyslexia' is not from lack of teaching. The person may gain insight by discussing his or her problem with fellow sufferers, and by finding out about past dyslexics such as Leonardo da Vinci. Special educational programmes are available for addressing dyslexic problems, as the phonetic approach to learning to read usually presents problems. NB: 'wait-and-see' is not wise—studies tend to find best results if identification is pre-school. Then exercises in sound categorization using rhyme and alliteration, with special teaching of letter sounds can be started early. medline:7528582

Doctors may be asked about dietary supplements with long-chain fatty acids (eg Efalex®). These do play a rôle in neurodevelopment, but evidence of benefit in dyslexia from large randomized trials is lacking. They do appear to reduce dyslexia-associated delay in dark adaption. medline:10617990

Compulsory hospitalization

► The patient must have a mental disorder, ie mental illness (p 314), mental impairment (p 314) or psychopathy (p 364), and need detention for treatment of it, or to protect himself or others, before compulsion may be used (if voluntary means have failed).

Admission for assessment (Mental Health Act 1983, section 2)

- The period of assessment (and treatment) lapses after 28 days.
- Patient's appeals must be sent within 14 days to the mental health tribunal (composed of a doctor, lay person, and lawyer).
- An approved social worker (or the nearest relative) makes the application on the recommendation of 2 doctors (not from the same hospital), one of whom is 'approved' under the Act (in practice a psychiatric consultant or senior registrar). The other doctor should ideally know the patient in a professional capacity. If this is not possible, the Code of Practice recommends that the second doctor should be an 'approved' doctor.

Section 3: admission for treatment (for ≤ 6 months)

- The exact mental disorder must be stated.
- Detention is renewable for a further 6 months (annually thereafter).
- 2 doctors must sign the appropriate forms and know why treatment in the community is contraindicated. They must have seen the patient within 24h. They must state that treatment is likely to benefit the patient, or prevent deterioration; or that it is necessary for the health or safety of the patient or the protection of others.

Section 4: emergency treatment (for ≤ 72 h)

- The admission to hospital must be an urgent necessity.
- May be used if admission under section 2 would cause undesirable delay (admission must follow the recommendation rapidly).
- An approved social worker or the nearest relative makes the application after recommendation from one doctor (eg the GP).
- The GP should keep a supply of the relevant forms, as the social worker may be unobtainable (eg with another emergency).
- It is usually converted to a section 2 on arrival in hospital, following the recommendation of the duty psychiatrist. If the second recommendation is not completed, the patient should be discharged as soon as the decision not to is made. The Section should not be allowed to lapse.

Detention of a patient already in hospital: section 5(2) (≤ 72 h)

- The doctor in charge (or, if a consultant psychiatrist, his or her deputy, eg a senior house officer) applies to the hospital administrator, day or night.
- A patient in an A&E department is not in a ward, so cannot be detained under this section. Common law is all that is available, to provide temporary restraint 'on a lunatic who has run amok and is a manifest danger either to himself or to others'¹ while awaiting an assessment by a psychiatrist.²
- Plan where the patient is to go before the 72h has elapsed, eg by liaising with psychiatrists for admission under section 2.

Nurses' holding powers: section 5(4) (for ≤ 6 h)

- Any authorized psychiatric nurse may forcibly detain a voluntary 'mental' patient who is taking his own discharge against advice, if such a discharge would be likely to involve serious harm to the patient (eg suicide) or others.
- During the 6h the nurse must find the necessary personnel to sign a section 5 application or allow the patient's discharge.

- Section 7: application for guardianship** Enables patients to receive community care if it cannot be provided without using compulsory powers.
- Application is made by an 'approved social worker (ASP)' or 'nearest relative' and also needs two medical recommendations.
 - The guardian, usually a social worker, can require the patient to live in a specified place, to attend at specified places for treatment and to allow authorized persons access.

Renewal of compulsory detention in hospital: section 20(4)

- The patient continues to suffer from a mental disorder and would benefit from continued hospital treatment.
- Further admission is needed for the health or safety of the patient—which cannot be achieved except by forced detention.

Section 25: supervised discharge This is as a result of The Mental Health (Patients in the Community) Act 1995, which has been incorporated within the 1983 Act.

- It allows formal supervision to ensure that a patient who has been detained for treatment under the Act receives follow up care.
- The application is made at the time of detention for treatment by the Resident Medical Officer. It is supported by an ASW and a doctor involved in the patients treatment in the community.
- A supervisor is appointed who can convey the patient to a place where treatment is given.

Section 117: Aftercare & the Care Programme Approach (CPA)

Section 117 requires provision of after-care for patients who have been detained on the 'long sections' (3, 37, 47, or 48). The CPA is not part of the Act but stipulates that no patient should be discharged without planned after-care: the systematic assessment of health and social needs, an agreed care plan, the allocation of a keyworker, and regular reviews of progress.

Section 136 (for $\geq 72h$) allows police to arrest a person 'in a place to which the public have access' and who is believed to be suffering from a mental disorder. The patient must be conveyed to a 'place of safety' (usually a designated A&E department or police station) for assessment by a doctor (usually a psychiatrist) and an approved social worker. The patient must be discharged after assessment or detained under section 2 or 3.

Section 135 This empowers an approved social worker who believes that someone is being ill-treated or is neglecting himself to apply to a magistrate to search for and admit such patients. The ASW or a registered medical practitioner must accompany the police.

*This Act operates in England; arrangements in Scotland are slightly different. ➤ The situation in the UK is changing and these pages should be read along with current legislation in the area where you are practising.

1 Lord Justice Keith, 1988 *The Times* May 28 2 R Jones 1991 *Mental Health Act Manual*, 3e, London, Sweet and Maxwell

Consent to treatment (Mental Health Act)

Emergency treatment to save life or to prevent serious harm to the patient can (must) always be given, overriding all the safeguards below, if the patient is unable to consent to treatment (eg owing to drunkenness).

Basic principle: In general a patient must give consent for any procedure (any touching), otherwise the doctor is liable under the UK common law of battery. This means that a competent person can refuse any treatment, however dire the consequences. Within common law there are defences to battery other than consent: if a patient is not competent to give or withhold consent, and if treatment is in their best interests, then treatment may be undertaken under 'necessity'. The defence of 'emergency' is to allow restraint where you must act quickly to prevent the patient from harming themselves or others (or committing a crime). For example, you could restrain a patient running amok on a ward before you have a chance to assess the situation. The Mental Health Act enables treatment of someone suffering from a mental disorder *for that mental disorder* under certain circumstances (p 398) and sets down some conditions for consent for treating in certain circumstances, as follows.

Consent is required and a second opinion is needed (section 57) All forms of psychosurgery require this level of consent. If patients are incapable of giving informed consent or decline consent, no treatment can be given.

The Mental Health Act Commission must validate the informed consent. It does this by sending 3 members to interview the patient and review the notes. One member is a doctor (who decides if the treatment is appropriate); others are non-medical. The patient must understand the nature, purpose, and likely effects of the recommended treatment for the conditions of informed consent to hold. If the patient is confused, demented, or would consent to *any* treatment, consent is not informed. Signing of an ordinary consent form is needed as well as the certificate from the Mental Health Act Commission. Although grades of informed consent are probably more realistic, the law is black and white: either the consent is informed, or it is not.

Consent is required and a second opinion is needed (section 58)

Note: no treatment can be given to a voluntary patient without his informed consent. If the responsible medical officer (RMO) feels that electroconvulsive or drug treatment for ≥ 3 months is necessary, and the patient is incapable of giving informed consent, or withholds consent, these treatments may be given provided a second opinion from the Mental Health Commission states that the treatment is necessary. Section 3 of the Mental Health Act is then used. The advantage of doing this is that the patient then acquires well-defined rights for review and appeal. If informed consent is given, the RMO must sign Form 38 (as well as the ordinary consent form) stating that the patient understands the purpose, need for, and likely effects of the treatment.

Children under 16 require the same standard of informed consent from a parent or guardian for prolonged psychotropic treatment. If parents are unwilling or unable to consent, a Care Order may be necessary—under which the local authority social services department takes over responsibility for the child. It must be shown that the child's proper development or health is being avoidably prevented or neglected; or that he is exposed to a moral danger; or that he is beyond the control of his parent or guardian.

Withdrawal of consent: Treatment must cease immediately, unless it is necessary to save life.

Seasonal affective disorder (SAD)

Lying thus in the sun one is liberated from doubts and from misgivings; it is not that problems and difficulties are resolved, it is that they are banished. The sun's radiation penetrates the mind as well as the body, anaesthetizing thought. AE Ellis 1958 *The Rock*

Some patients find that symptoms of depression start in the winter months, and remit in spring or summer. It has been postulated that disordered secretion of the indole melatonin from the pineal gland is to blame in some of these patients with 'SAD'. Melatonin, the hormone of darkness, is secreted by the pineal only at night—eg at 30µg/night; evidence supporting the rôle of melatonin in depression includes the following:

- The antidepressant effect of light is potentiated by early-morning administration in circadian time, optimally at ~8.5 hours after melatonin onset or 2.5 hours after a sleep's midpoint. medline:11146760
- A dose-response effect exists between the amount of light administered in phototherapy and the degree of improvement in depression (as measured on Hamilton ratings). 6h/day of increased light brought about a 53% decrease in scores, whereas treatments of 2h (or red-light treatment) produced only a 25% reduction. These effects were correlated with suppressed plasma melatonin concentrations at 23.00h. Variables of uncertain importance include: the type of light (device & spectrum); distance between patient and source; and the duration of treatment. medline:10353446

NB: therapy should stimulate the nasal retina, as retinal ganglion cells projecting to suprachiasmatic nuclei are unequally distributed. medline:10194647

However, evidence in this area is often contradictory, and it is probably unwise to rush into recommending light for all patients whose recurrent depressions start in the autumn or winter. This might have the undesirable effect of enticing such patients to book unaffordable winter holidays to exotic locations—with inevitable disappointments and recriminations.

Community care

Since the early 1980s, most UK inpatients with mental illness have had the focus of their care moved from hospital to the community. The aim has been to save money and improve care: but, in the UK, this policy is now being partly reversed. Has community care failed, or have there been successes?

Five questions keep recurring, each (ominously) prefixed by a 'Surely. . .'

In general, the problem is not the severity of the mental illness, but the social context in which it occurs, which determines if community care is appropriate.

Not so—at least not necessarily so. Some concentration of resources can take place in the community, for example, in day hospitals and mental illness hostels (p 326–7). It is also true that the 'bed and breakfast' element of inpatient care is likely to be quite expensive, particularly if the running and maintenance costs associated with deploying inpatient psychiatric services are taken into account. In most studies, the cost of each type of service doesn't differ much, and sometimes good community care turns out to be cheaper. medline:10579701 & 9596049

It is now clear that however careful selection of patients for community care may be, this will still be a very serious problem.

The implication here is that the skills available in bed-and-breakfast accommodation may be inadequate at times of day when there is no other support, other than the general practitioner. Studies that have looked at this have certainly found an increase in non-hospital residential care in those selected for community care, and this increase may be as much as 280% over 5 yrs. In the UK, new proposals guarantee 24-hour open access to skilled help, but it is not known what pressures this will put services under.

This will be so if every patient has a lengthy care plan, and repeated risk assessments. Concern for safety may also spawn a non-therapeutic custodial relationship between health-worker and patient.

Advantages reported for community care are: better social functioning, satisfaction with life, employment, and drug compliance—but in randomized studies in the UK these advantages are not always manifest. Furthermore, trends have been repeatedly found indicating that the longer studies go on for, the harder it is to maintain the initial advantages of community care. If it is hard for teams to keep up their enthusiasm during a trial, it will probably be even harder when the trial period has ended, or when team members are ill. These constraints may in part explain the observation that with inadequately funded and supervised community care, patients can fail to get essential services, and when hospitals are being run down, and a patient's condition worsens, so that 'sectioning' followed by admission becomes impossible, the patient is left in the community 'rotting with his rights on'.

& is one way out of this impasse (here a key-worker has direct responsibility for care plans). This set-up helps ensure more people remain in contact with psychiatric services (NNT = 15); this inevitably increases hospital admission rates. medline:10796288 & 10796415

Poverty and mental health

Social deprivation is positively associated with premature mortality, and poverty makes almost *all* diseases more likely (but not Hodgkin's disease, eczema, bulimia, or melanoma). medline:10752704 See *Health and social class*, p 463. In the uk, the number of homeless people is 1–2 million. >30% suffer from mental illness (10% have schizophrenia), most do not know where to go for help, and most have no doctor. A 3-tier strategy may help.
1 Emergency shelters 2 Transitional accommodation 3 Long-term housing

The cost in health terms to society and the individual is enormous. Diseases and symptoms such as diarrhoea, which may pass as a minor inconvenience to the well-housed, may be a major hurdle for the homeless, with severe social and psychological effects. Capture-recapture techniques show that the *unobserved* population of the homeless is about twice that observed. This method of enumeration collects samples (lists) and looks for tags (duplicates) in subsequent counts, and from this determines the degree of under-counting. If all in the subsequent count are duplicates, then there is no underestimate of the original count. Statistical techniques can allow for migration in and out of the population area. medline:8298348 These studies show that psychiatric morbidity is greatest in the observed homeless populations: the implication is that the psychiatric illness makes these people more 'visible'.

In the uk, as in many other Western countries, what started out as an enlightened policy of looking after people with mental health problems in the community has resulted in large numbers of psychiatric patients living on the street in great poverty—relieved by occasional admissions to acute units. This 'revolving door' model of care has failed many patients, not least because continuity of care has been compromised.

One way to stop poverty is to pay people not to be poor—specifically not to have more than 1–2 children. In the early 1990s, the Singapore government offered poor families ^{SP}\$26,400 over 20yrs if they have ≤ 2 children.

In the uk the 'poverty line' weekly income is calculated on an individual basis depending on circumstances, so for an unmarried mother with 2 young children the figure is £93.85. In fact, state benefits may not amount to this sum. She will receive Income support personal allowance of £46.50 plus £15.95 per child, plus £10.25 family premium with a £5.20 single parent premium (total £93.85). Further benefits such as child benefit or maintenance will lead to a proportionate reduction in this total. From this she might typically pay £3 for water bills, £2.20 for council tax, and £10/week for fuel (in winter). In council flats there are no furnishings. Grants (eg £500) are scarce, so DoH can make a loan for this, deducted at source (eg £13/week)—so the amount some families are living on is less than £60. NB: these figures are from the mid-1990s; valuations change frequently (ask at freefone 0800 882200).

Note that there is no evidence that simply living in a deprived area makes a person more prone to illness and death. All the excess mortality and morbidity is explained by the person being poor.

medline:7804047

Their immediate neighbours who are not poor do not share the same risk. The implication is that targeted health care should be directed towards poor people *wherever* they live, not at poor areas.

Postnatal depression

■ *Suicide is a leading cause of maternal mortality.* The psychiatric and psychological phenomena of pregnancy and birth include the very common, and usually mild, 'blues'; puerperal psychosis; and postnatal depression (the risk of developing this latter is 3-fold that of those with no recent pregnancy). There are many causes: individual and social circumstances; genetic; hormonal change (see below).

■ *Natural history* Reviews state that most postnatal depression resolves within 6 months, so we find ourselves thinking: 'Good! Those trusty psychiatric tools of "wait-and-see", and the passage of time will be my main tools in this illness: nothing depends critically on what I do now, to this poor depressed new mother with whom I am now confronted'. NOT SO!

■ *Consider these facts* For the patient, 6 months is a long, long time.

- For the infant, 6 months is more than a long time: it's literally an age.
- Suicide is a waste, but for a young family, a mother's suicide is especially destructive—unthinkable, indeed, for those who have not experienced it.
- There is impaired cognition and social functioning in the offspring of mothers with postnatal depression.

medline:9489170 & 8163630

We therefore submit that our actions may spawn critical outcomes. So what should our actions be? The first step is to try not to be caught unawares by a major depression that apparently strikes like a bolt from the blue, but which, in reality, has been building up over time. Pregnancy and infant-motherhood is supposed to be a time of unclouded joy. We professionals often collude with this view. We are always hearing ourselves saying: 'Oh Mrs Salt, what a lovely baby! You must be so pleased—and you always wanted a little boy, didn't you? We are so delighted for you. . . ' But what if *she* is not delighted? She hardly dares confess her traitorous thoughts that she is unaccountably sad, that she spends the nights crying, and that her exhausted days are filled with a sense of foreboding that she or some other agency will harm the child. The place to start to pre-empt these feelings is in the antenatal clinic. By addressing these issues, and, later, in the puerperium, by expressly asking after them, you give permission for the new mother to tell her woe. When this is revealed, counselling, and input from a health visitor and a psychiatrist is appropriate, as is particularly close follow-up. You may need to arrange emergency admission under the Mental Health Act: but the whole point of being prepared for postnatal depression is to avoid things getting this bad.

■ *Pharmacology* is as effective as cognitive-behavioural counselling in the short-term. More trials with longer follow-up are needed to compare antidepressants and social interventions.

medline:11406023

Although all antidepressants are excreted in breast milk, tricyclics and SSRIs are rarely detectable by standard tests. Observe babies for possible SES; it may be best to stop breast feeding if large doses are used.

Adding (p 354) may help (ECT may also be needed). Trials show that is partly effective in severe postnatal depression.

The Edinburgh Postnatal Depression Scale (EPDS, OPPOSITE) was used to assess outcome (along with other measures). In one study, the score started off at 21.8, falling within a month to 13.3—taking some patients out of the 'major depression' category. The control group also improved over time (from 21.3 to 16.5)—but still remained severely depressed. Dose régime: 3 months of transdermal 17 β -(o)estradiol (200 μ g/day) for 3 months on its own, then with added dydrogesterone 10mg/day for 12 days each month for 3 more months. CI: uterine, cervical, or breast neoplasia; past thrombo-embolism/thrombophlebitis; breast-feeding.

medline:10796270

Please underline what comes closest to how you have felt in the last 7 days.

- 1 I've been able to laugh & see the funny side of things: as much as always could
not quite so much now
definitely not so much
now not at all
- 2 I've looked forward with enjoyment to things: as much as I ever did
rather less than I used to
definitely less than before
hardly at all
- 3* I've blamed myself unnecessarily when things went wrong: yes, most of the time
yes, some of the time
not very often
no, never
- 4 I've been anxious or worried for no good reason: no, not at all
hardly ever
yes, sometimes
yes, very often
- 5* I've felt scared or panicky for no very good reason: yes, quite a lot
yes, sometimes
no, not much
no, not at all
- 6* Things have been getting on top of me: yes, most of the time I haven't been able to cope at all
yes, sometimes I haven't been coping as well as usual
no, most of the time I have coped quite well
no, I have been coping as well as ever
- 7* I've been so unhappy that it is difficult to sleep: yes, most of the time
yes, sometimes
not very often
no, not at all
- 8* I've felt sad or miserable: yes, most of the time
yes, quite often
not very often
no, not at all
- 9* I've been so unhappy that I've been crying: yes, most of the time
yes, quite often
only occasionally
no, never
- 10* Thoughts of harming myself have occurred to me: yes, quite often
sometimes
hardly ever
never

Answers are scored 0, 1, 2, and 3 according to increased severity.
*Some are reverse scored (3, 2, 1, 0). Add scores for 1–10 for the total.
Let her complete the scale herself, unless she has limited English or has difficulty with reading. EPDS may be used at 6–8 weeks to screen postnatal women, eg at the postnatal check-up. A score of 12/30 has a sensitivity of 77% and a specificity of 93% for postnatal depression.

The main point is that it's all too easy for doctors and midwives to go on thinking 'it's just postnatal blues' when in fact a serious depression is unfolding. The above tool is helpful in making this distinction.

5. Primary care

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Universal patient orientation – Every page in all chapters. All diseases are relevant. This is a key feature of primary care which is the axis around which all medical events revolve, every day drawing millions of patients to it before scattering a few to every corner of the medical world. Only in primary care is the full gamut of what can go wrong with people's lives made manifest. Because of the huge and undefined range of general practice, it has been said (not just by GPs) that the general practitioner 'needs to be the most comprehensively educated of any kind of doctor.'⁶

Primary care is not a specialty in the usual sense, and GPs make much of being the only remaining generalists (they were also the first). They are specialists in their patients, rather than in disease—having no special and unique knowledge of disease (except for disease in its earliest stage), and having no unique treatments. They can answer questions like ‘Why doesn’t Miss Phelps ever attend the antenatal clinic? What would have to change for her to do so?’ There are few medical instances in which GPs are in a position to offer more skill and expertise than all other doctors—but there are many circumstances when the GP’s understanding of his patient is what counts, and for which no amount of expertise can be a substitute. So when, for example, Miss Phelps’s baby dies, to whom does she turn in her distress? She is not a medical problem; she is not an obstetric problem; she is not even a psychiatric problem—she is Miss Phelps. And the doctor who specializes in Miss Phelps is (or ought to be) her GP. He is the one who knows what to do and what to say; when to be quiet and when to explain; and when to appear and when to disappear, humbled as he is by having witnessed a wealth of such grief.

With knowledge of the patient, and knowledge of disease in its earliest phase, there is a third skill which is vital: the ability to diagnose health taking into account all the definitions of health on p 414. If a patient is asking ‘Am I well, or could I have cancer’ the doctor who is always responding by saying ‘... Well, I’m not sure. ... let’s do one more test. ...’ is sometimes a bad doctor, and always an expensive one. Some doctors have the gift of appraising a patient, and knowing almost instinctively whether to treat now, or investigate, or refer, or reassure. Nowhere is this skill more apparent than in general practice—and this instinctive skill is born of countless contacts with patients from all walks of life, with diseases affecting any organ of the body or mind. A chest physician may tell you that your lungs are healthy, but if, on your way out, you say ‘Oh, and I’ve also got this pain in my leg. ...’—then he may feel out of his territory. He thought he was sending his patient away rejoicing in his health, but then one new symptom undermines this, and he is likely to end up saying ‘Go and see your GP’. It is then up to the GP to reassure, treat, or to orchestrate the deployment of all the many sophistications of technological medicine.

Even if all our scientists had unlimited abilities and facilities, there would still be a limit to technological medicine. Even if we could turn the whole world into a hospital so skilled as to be able to manipulate the totality of the *milieu intérieur* there would still be a vast abyss between the possibility of health and its attainment. On the other side of the abyss is the patient, surrounded by unknown (but not unknowable) myths, hopes, fears, ideas, expectations, and fantasies directing him away from the very things that could help him most. It is the GP’s rôle to bridge this abyss, not so much to haul the patient to safety, but rather to be the platform on which the patient crosses of his own (enlightened) free will. In bridging this gap, the GP needs sure foundations on both of its sides, and this is the great challenge of general practice—to foster in oneself an equal love of, and an excellence in, both the technological and the personal realm.

Although general practice is not a specialty in the usual sense, it is a discipline, as it creates and publishes original material about its sphere of activity in peer-reviewed journals (eg *British Journal of General Practice* and *Journal of Family Practice*, which are in the top 5 world-class medical journals related to a specific discipline, as ranked by citations and immediacy). Because these journals are the lifeblood at the scientific heart of the discipline, they are not necessarily an easy read—but they can repay careful study.

Giving yourself to the highest bidder?

*'A healthy man is, above all, a man of this earth, and he must, therefore, only live the life of this earth for the sake of order and completeness. But as soon as he falls ill, as soon as the normal earthly order of his organism is disturbed, the possibility of another world begins to become more apparent, and the more ill he is, the more closely does he come into touch with the other world.'**

We do not always understand the worlds inhabited by our patients but because of this, do not assume that our patients must travel these worlds alone. Taking time to find out what it's like for our patients is the first step in forging an enduring doctor–patient relationship. When our patients know that we are travelling with them, and that we will not abandon them, they will accept our foibles, even our errors. But how can we cope with this big commitment—big enough for one person, let alone a few thousand of our dependent patients? How can we do all this without destroying ourselves? Do we give in to the highest bidder? Here are some insights from a woman in a crisis with too many conflicting rôles: daughter, mother, lover, and so on: 'There is a battle going on for my soul. . . and I cannot just give it to the highest bidder. I have an interest in it too. . . I have a duty to many people and somehow I will discharge it. I have a duty also to some continuing part of myself. I have . . . ripped open my self-protective layers. I see now what I am. It's not a question of "happiness". I don't value my own more—or much less than anyone else's. It's something more lasting: it's a question of being faithful to an essence.'**

If we spend day after day in surgery without attending to our other rôles we are not necessarily better than a person who leaves work on time, so enabling a visit to a grandparent, or a dialogue with a daughter, or time for recreation. The medical world encourages the dangerous delusion that we are somehow inadequate if we do not give our all. What gives rise to this is the delusion that the best unit of measurement of our medical lives is the single consultation. This is how we are assessed, as if there were no valid distractions during consultations—as if our own needs were non-existent, and 100% of the focus is placed on the patient sitting in front of us. But what if you should not really be seeing this patient at all, but should be out on a visit which might or might not be urgent? Or would it be better to be on the phone, talking to a possibly suicidal patient who has missed their appointment? Perhaps you need to do all three. Then you will do none of them well. If we are going to be successful in primary care, with its unending responsibilities, we have to recognise that the best doctors may not do anything very well. The best doctors just make the least bad decisions on how to spend their time, and themselves. As with the woman above, they do not simply give their soul to the highest bidder. Don't feel guilty about this. To give yourself to the highest bidder would be a betrayal: not even saints do this.

*Dostoyevsky *Crime And Punishment* Tr David Magashack Penguin, p 305 <http://www.bartleby.com/318/41.html#78>

** Sebastian Faulks 2001 *On Green Dolphin Street*, Hutchinson, p 239

► If you speak the language, you may eventually come to understand it.

ACBS Advisory committee on borderline substances; prescriptions thus endorsed reclassify special foods (eg without gluten) as a free drug

BMA British medical association—a kind of trade union for doctors, which also convenes various patient-related enterprises. Its organ is the *BMJ*

CHI Commission for health improvement—aims to improve standards by: assessing every NHS organization and making its findings public—investigating when there is serious failure—and checking that the NHS is following national guidelines. It advises on best practice www.chi.nhs.uk

CPN Community psychiatric nurse

DH/DSS Government department of health/department of social services

DNA Did not attend (for a booked appointment ≈ waste of NHS resources)

EBM Evidence-based medicine, or the journal of the same name

ECR Extracontractual referral; ie no existing contract exists (∴ costly)

EHF Electronic health record accessible anywhere in the NHS; doesn't exist

FMed3/5 Forms for sick pay, p 421

FP10 The form on which NHS prescriptions are written—ie scrip(t)s

GMC General medical council

HIMP Health improvement & modernization program, eg ↓ smoking rates

IM&T Information management and technology

IS Local implementation strategy, eg for IM&T

LMC Local medical committee (which approves or withholds approval for various centrally promulgated non-statutory health policies)

MASTA Medical advisory service for travellers abroad. www.masta.org

MDU/MPS Medical defence union and the medical protection society

MESH Medical subheadings used in medline searches, see p 447

NELH National electronic library for health www.nelh.nhs.uk

NHS National health service: a system for providing health services universally free at the point of use, funded out of general taxation

NICE National institute of clinical excellence

NMC Nursing and midwifery council (replaces UKCC)

NPSA National patient safety agency for reporting critical incidents

NSF National service framework (eg for diabetes, heart disease etc, p 463)

OCM Oxford Clinical Mentor: decision support www.mentor-update.com

OTC Over the counter medicine

OXCHECK Oxford study on prevention. <http://bmj.com/cgi/content/full/310/6987/1099>

PCO Primary care organization (usually a Trust—PCT)

PACT Trademark of the prescription pricing authority (PPA)

PGEA Postgraduate educational allowance (eg PGEA-approved study session)

POM Prescription-only medicine

PRECE Primary care clinical effectiveness: an exacting standard of prevention involving coronary heart disease, diabetes, etc.) medline:11467271

PRODIGY Prescribing advice triggered by entering Read codes

RCT Randomized controlled trial

SFF Service and financial framework—a financial plan

SLC Selected list scheme; written on an **FP10**, this makes Viagra etc free, eg if DM, MS, parkinsonism, prostate ca, spina bifida, cord injury or post-polio

SMR Standardized mortality ratios

SPN Supplementary prescribing nurse (p 418)

TQM Total quality management www.euro.found.ie/1997/05/Feature/UK9705113E.html

DVLA Driving vehicle licensing authority www.dvla.gov.uk/ac_a_glance/content.htm

Some definitions and measures of health

Primary care and distributive justice The who Alma-Ata statement declares that primary care should 'be made universally accessible to individuals and families in the community, by means acceptable to them, through their full participation, and at the cost that the community and country can afford to maintain in the spirit of self-reliance. . . [and] addresses the main health problems in the community, providing promotive, preventative, curative and rehabilitative services accordingly.' Factors affecting access to health include finance, ideology, and education.

- 4 or job descriptions**
- To clear the waiting room efficiently (kindly if possible).
 - To deal with the realities of patients as you find them (eg loneliness, addiction, poverty, and mental ill-health) rather than ideal patients (eg interesting diagnostic problems leading to technically brilliant cures).
 - To be skilled in: prioritization; delegation; health-need measurement; rationing; purchasing/delivery of primary healthcare; time- and staff-management.
 - To give healthcare to people irrespective of age, sex, illness, or social status.
- To make early diagnoses, framed in physical, psychological, and social terms.
 To make initial decisions about all problems presented or unearthed.
 To arrange continuing management of chronic, recurrent, or terminal illness.
 To practise in co-operation with other colleagues, medical and nonmedical.
 To treat, prevent, and educate to promote health of patients and their families, while reconciling professional responsibility to the community.

Health 4 definitions to compare: Health is: I The absence of disease.

II A state of complete physical, mental, and social wellbeing. who definition

III A process of adaptation, to changing environments, to growing up and ageing, to healing when damaged, to suffering, and death. Health embraces the future so includes anguish and the inner resources to live with it. I Illich, 1976

IV Any process enabling the giving or promoting of life.

V 'Health is whatever works, and for as long'. J Stone, 1980, In All This Rain

All the above have limitations, but I and IV seem least counter-intuitive. Consider the following: • Was Charles I healthy as he laid his head on the executioner's block? • What about a priest in the act of losing his celibacy? • Can a heart with a prosthetic valve which is gradually wearing out be healthy? • Was Gandhi healthy at the end of a hunger strike? • Can animals or babies be healthy? • What about death in childbirth? 'Answers' below:

Healthy according to definition:

	I	II	III	IV	V
King Charles on the scaffold	yes	no	yes	yes	no
Fasting Gandhi	yes	no	yes	yes	yes
Babies and animals	yes	yes	no	yes	yes
Heart with failing valve	no	no	no	yes	yes
Priest losing his celibacy	yes	no	yes	yes	yes
Death in childbirth	no	no	yes	yes	yes

Measures of health Scores on the health survey Short Form 36 (SF36) are reproducible and quantifiable, related to patients' clinical state, the GP's decision to refer, and GP's views on severity. It is valid when combined with a patient-generated index of quality of life (which asks patients to name the 5 most important activities/areas affected by their condition, and to value importance of improvements to them) and a daily time trade-off calculation (how much time would you give up to be in perfect health?). By combining instruments, defects in one can be mitigated (eg the SF36 asks if health limits your ability to walk a mile—irrelevant if the patient does not need or want to walk much). ¹ is the difference between the state now and a goal. ² Needs may be ranked by the distances between states and goals.

What are the determinants of health?

The answer is **wealth**. With wealth comes more stable political systems, and these are what are necessary for literacy and education to flourish, which in turn leads to easy access to clean water (the key issue, as more than 1 billion people have no access to clean water), and the possibility of developing equitable health delivery systems.

A consensus is emerging in some public health units in the UK on what have been the most important influences on health in the last 20 years.

- Improvements in living standards from greater disposable income.
- Improvements in the environment, eg housing and road safety.
- Reduction in tobacco use (but the young continue to take up smoking).
- Improvement in education and communication.
- Better, more effective health services, with easier access to them.

Other influences include nutritional improvement, smaller families, exercise, community self-help, and the prevention of HIV infection.

Future determinants of health are thought to rest on:

- Reducing inequalities in access to health care, and in its content.
- Reducing unemployment (a potent cause of ill health, see p 406).
- Decline in tobacco consumption in all age groups.
- Better health services with more effective, more acceptable treatments.
- Fewer underdoctored areas (currently defined as populations where there are fewer than 52.695 GPs per 100,000—ie a list size of >1898 per whole-time GP)—and more GPs in deprived areas. Funding more GPs has been calculated as one of the most efficient ways of reducing mortality.
- Education capable of influencing behaviour so that exposure to identified risk factors is reduced.

Better protection of the environment and better housing.

- Applying existing knowledge in a better, more systematic, way.
- Introducing a more patient-centred style of health care, in which patients are not the passive recipients of care, but well-educated partners in the struggle against disease and its causes.

Primary health care

Primary care is the system of care used by people as their first contact with health services. Where needed, referrals are made (eg in ~10% of UK patients) to centres of secondary care—usually district hospitals. Further referral may be necessary to regional ‘super-specialists’, so constituting tertiary medical care. This seductively simple model of medical provision misses out entirely on the cornerstone of primary health care—the responsibilities that individuals and families have for their own physical and mental wellbeing. *Up to 90% of all health problems are taken care of outside the official health care system.* Unless individuals and families act on their own initiative to promote their health, no amount of medical care is going to make them healthy; coronary artery bypass grafting may be helpful, but it is no substitute for families taking the initiative in promoting their own health by trying to live sensibly (eg *vis-à-vis* diet and smoking) and by learning to look after their own health. In fostering and enlightening this sense of initiative, a large number of groups are involved—not just doctors but also teachers, health educators, politicians and, above all, the families themselves. So in assessing how good a community is at primary health care, one needs to look not just at medical care, but also at social, political, and cultural aspects as well. One must ask questions such as: *Is society making it easy for individuals to choose a healthy lifestyle?* and *How is society promulgating health education?* and *When does this education become indoctrination?*

Primary health care defined as a set of activities

- Providing safe food and water
- Ensuring freedom from want.
- The basic treatment of illness
- Provision of necessary drugs.
- Preventive care (p 426)
- Enabling Maslow's hierarchy (p 315)

Primary health care defined as a strategy No country in the world is rich enough to provide its citizens with everything that medical care can offer. This fact pinpoints the need for the efficient use of limited resources—and this presupposes an effective system of primary health care. To be effective, this must be accessible; relevant to the population's needs; properly integrated; have full community participation; be cost-effective, and characterized by collaboration between sectors of society.

Some UK based facts ● GP services are <10% of total NHS costs.

- 90% of illness known to the NHS are handled in general practice.
- The number of unrestricted principals in the UK is ~40,000 with ~2000 GP registrars, and 1000 assistants. Supporting staff: ~2/GP. www.rcgp.org.uk

Intermediate care

medline:12052787

This newly named type of care lies between traditional primary care and secondary care. It integrates facilities from many areas to address complex health needs which do not require use of district general hospital services. Examples include pre-admission assessment units; early and supported discharge schemes; community (cottage) hospitals; domiciliary stroke units; hospitals-at-home schemes; rehabilitation units. It is one of the mechanisms by which health and social services mesh to allow patients to receive the most appropriate care. Its main advantages are that it allows:

- 1 Effective care, close to home.
- 2 Best use of new technologies, such as information technology, near-patient testing, and phone-activated devices to summon help.
- 3 Cost-effective use of resources.
- 4 Less rigidly demarcated professional rôles.
- 5 Creative integration of working practices.

Don't think of it simply as reducing bed-blocking, but it does! medline:12038232

Self-care and empowered self-care

Simple self-care constitutes the health activities which we do on our own and within families—such as brushing our teeth, or going to bed with aspirins during 'flu. Empowered self-care is what can happen when primary, secondary (distric general hospital), and tertiary care (eg regional burns units and cancer specialisms) work together with social services within the context of the family life cycle. Crucially, it uses the principles of intermediate care (see opposite). • *Of any health care system, ask how rich and deep are its community roots?* How many options are there for the care of this sick old lady who has a bad chest and is temporarily off her feet? If your health care system lacks depth, and if ties of religion and family are loose, the only option may be an emergency admission to a high-technology hospital. Emergency admissions in the UK and many other areas have been climbing inexorably—for many reasons. One important reason is lack of options at the primary care level.

Whenever you think of admitting a patient to hospital, ask *When are the other options?* Do this not just to save the hospital work, but to force you to find out what your patient really wants, and to ensure that the most appropriate level of care is found. Examples of options are:

Neighbourly help	Hospice; sheltered housing	Hospital at home
Nursing/rest home	Social services-run home	Twilight nurses at home
Domiciliary physio	Occupational therapy	Fast-response nursing

When you think of these options, don't think *structure of services?*—think *universal health workers*. Universal health workers have various skills: find out about them, and judge them not according to historic professional codes but according to how good they are at empowering self-care. No health service can look after most patients most of the time. Empowered self-care in the context of interdependent social and medical services is not some new option that may or may not be used: it is the *only* option for health services which aim to look after more than one patient. Without this idea of empowered self-care hospitals become places of passive dependency, they get too full, and you cannot get people in, and you cannot get people out. NB: if you think that empowered self-care is a cliché, try doing your diabetic clinic without it: you will always fail.

Empowered self-care entails choice (p 171 & p 315), dialogue (p 368 & p 458), knowledge of mental states (p 324) informed consent (p 762 & p 168), literacy/education (p 438), participation in planning and respect by professionals for lifeworlds other than their own (p 321). We have to harmonize our care-plans with the patient's belief system. So if, for example, a man takes strength from meditation, this should feed into the dialogues which inform his empowered self-care. This yields more patient satisfaction **and** improved outcomes: see p 458. medline:11443762 & 10146013 & 9179834

Barriers to this type of care

- People who are rendered helpless and hopeless by unemployment, poverty, and family strife. Others who have difficulty accessing care include: the homeless, refugees, drug abusers, ethnic minority groups, and patients living in rural areas without public transport.
- Professionals who want to monopolize and medicalize health.
- Nations which are keener to take up arms than to vaccinate them.
- A world which behaves as if it does not know the meaning of social justice and equality, and in which rich and poor fail to share common objectives—or simply fail to share anything.

The primary health care team

The antithesis is:

Nature favours the first maxim:

when we die all our tasks are either forgotten or delegated, often by default. So the question is not *whether* to delegate, but *when*, and *to whom*. The principle of team work is:

Understand the team members. Some may specialize eg in surgery, diabetes, endoscopy—or have a rôle in his or her Primary Care Organization (PCO) eg in commissioning hospital care; **Medical and other students** may also be present.

A good question for any partnership to ask itself is what is its range of skills, and is postgraduate training being arranged to fill in any lacunae? Subspecialisms need not undermine the central rôle of the GP as generalist.

Community nurse. (Employed in England by Primary Care Trusts.) Activities: post-op visits for dressings and the removal of sutures, dressing leg ulcers, and giving 'all care' to the elderly housebound, giving injections (eg to blind diabetic patients), and supplying incontinence and other aids ± catheterizations. **Nurse prescribers** have additional training (37 days) and are **SNPs** or **SNPs** (SNPs only prescribe according to protocols once a diagnosis has been made. medline:11865217

Midwives. They do antenatal classes, clinics, home visits, and home deliveries. They have a statutory obligation to visit in the puerperium for the 1st 10 days (she has right of access). At 10 days the health visitor takes over.

Health visitors have nursing and midwifery backgrounds, plus health visiting qualifications. Rôles: developmental testing of children; immunization advice; breast feeding; minor illness in children; handicap; advice to adults about diet & smoking; implementing health education officer (p 438) strategies; screening of the elderly in their homes; bereavement visits.

Practice nurses Activities include tests (urine, taking blood, audiometry, ECGs, peak flow); advice (eg diets); treatment, eg ear syringing, injections, *prevention* and *audit*—eg immunizations, BP, cervical smears, family planning (eg holding the English National Board Certificate); *chronic disease management*—diabetes/asthma clinic, IUCD insertion, chaperoning (usually she is too busy).

Nurse practitioners diagnose and initiate treatment. Patient satisfaction is high. No increase in adverse outcomes has been found; consultations are longer by 3–4 min, and more tests are done. medline:11934775 & 10764366

Counsellors. Up to 30% of primary care patients have psychological conditions requiring care, and to help with these, counsellors may be employed on site or remotely; outcomes are improved by this therapy. medline:11869583

Receptionists and secretaries. These are employed by GPs and salaries are partly reimbursed. Just because they are 'nonmedical' it does not follow that they do no medicine—eg a depressed patient may find quiet times to see a receptionist. Receptionists may be taught to take blood, test urines, do blood pressures, capillary glucose, ECGs, or audits. With the advent of e-mail referrals direct from the GP's desk, the rôle of secretaries is changing: she may now wield a sphygmomanometer, or do the audits.

Practice managers lead on finance, employment law, tax, risk assessment/reduction, health & safety, financial management, and clinical audit.

Others. Social worker, psychologist, facilitator (to encourage preventive activities), physiotherapist. NHS direct; NHS walk-in services; nurse-led personal medical services; community pharmacists, health education officers (p 438), community physicians.

Single-handed and small practices

In the UK, much of the progress in general practice over the recent past has evolved in the context of group practice and the primary health care team—the credo of modern-day doctoring. Single-handedness puts a question mark over the primacy and validity of these ideas. The benefits of highly ordered, management-led, high-investment, team-based general practice are real and not to be gainsaid; larger practices may also have easier rotas, with possible benefits to the mental health of GPs. But at what price have these benefits been bought? Can small be beautiful?—and what does this concept mean for health care the world over? Let us start with the observation that general practice is about people. Their illnesses are the incidental accidents, and their deaths the only certainties which mark the impingement of these individuals on our general practice nights and days. These contacts in the night are, for the larger practices, usually boring or worse. There is no before or after: just a job to be done. For the single-handed GP doing more of his or her own visiting, these occasions may also be boring—initially. But the utility and the interest inherent in these visits can, with continuity of care, stretch over the decades, as this example illustrates. One of us (JML) was called late to an unrousable, sweating 60-year-old man, whom we admitted to hospital with suspected septicaemia—which, years later, turned out to be the presenting symptom of an occult, indolent malignancy. We looked after him until his death at home. The continuity of care made the job interesting for us, not the clinical details. But the continuity does not stop at the end of one life. Now, whenever we see his wife, on however trivial or grave a problem, we have this shared bond. Recently she refused hospital admission for pneumonia, and the doctor could use this shared bond to induce her, over 24h, to change her mind. Of course, doctors in large groups will be able to tell of similar instances. Our point here is simply that these stories are more common, the more personal your care.

The dangers of exhaustion and isolation in single-handed practice are real, but they are usually pointed out by people who are thinking about isolation from one's colleagues. (How many partnerships are only partnerships on paper, and conceal or exemplify worse things than isolation?) The single-handed doctor is more likely to compensate for isolation from colleagues by identifying more with his or her patients.

As one GP has commented '... I am now about to start a day of single-handed general practice as my partner is away. The day is unplanned, with no appointments. People just turn up, bringing their infarcts, their sorrows, their trivia, and their life events to me. Some of the people who will come have not yet even fallen ill. There are the coronary artery plaques on the point of rupturing, the dizziness before the fall, the hallucination before its enactment, and someone is now writing a note to explain the impending suicide or the fact that they are leaving home forever, and taking the kids with them. All this is in the future; but for now, none of this has happened yet. I am completely up to date, and I command my general practice sitting behind an empty desk. I saw the last person who wanted to see me yesterday. There is no waiting list. Just the unknown. And what of the concept of the primary health care team? Can it really be called a team when I own the premises, hire most of the staff, pay them their wages, draw up their job description, and stipulate the bounds of their practice? It is certainly not an *equal* partnership. But when the patient's dizziness *does* lead to the fall in the village shop, as it did yesterday, I will be on hand to patch up the old lady, and the receptionist will kindly finish off the patient's shopping for her, and escort her home. We think that the price of a personal service is well worth paying.'

Time and the general practitioner

A typical GP with a list of ≤ 1900 patients of whom $\sim 15\%$ are over 65yrs might see 50 patients in a day with 5 home visits (mean=9745 consultations/GP/yr costing $\sim \pounds 13$ each). The major categories of care are for self-limiting illnesses (65%), chronic illness (20%), and acute major illness (15%). The most common conditions are listed below as rates/100,000 patients/yr derived from validated coded data in representative practices of a typical UK county (note that the idea of drawing health purchasing conclusions from some idealized 'typical' practice is flawed: there will be very few typical practices).

Infections/infestations	♂: 11,194	♀: 13,924
Neoplasms	♂: 2473	♀: 3695
Endocrine/nutrition/metabolic	♂: 3346	♀: 4326
Blood diseases	♂: 617	♀: 1609
Mental disorders	♂: 5440	♀: 8,442
Neurological/sense organ diseases	♂: 16,172	♀: 19,305
Circulatory diseases	♂: 10,359	♀: 12,902
Respiratory diseases	♂: 24,708	♀: 29,563
Digestive diseases	♂: 6806	♀: 8229
Genitourinary diseases	♂: 3657	♀: 15,654
Pregnancy/childbirth/puerperium	♂: 0	♀: 1467
Skin/subcutaneous diseases	♂: 12,653	♀: 15,817
Musculoskeletal/connective tissue	♂: 13,736	♀: 18,120
Congenital anomalies	♂: 472	♀: 444
Perinatal conditions	♂: 59	♀: 86
Symptoms/signs/ill-defined	♂: 25,719	♀: 38,373
Injury and poisoning	♂: 13,810	♀: 12,605

medline:8646146

Time and the consultation ▶ Does heavy demand produce short consultations, or do short consultations produce heavy demand by failing to meet patients' needs? GPs' average consultation time is ~ 7 min (with some consultations lasting $\sim 1/2$ h). This seems short, but remember that over a year the average time spent with each patient is nearer one hour. The consultation time influences the degree of patient satisfaction, and may influence the consultation rate (2.5–6/patient/yr), with lower return visit rates for longer consultations, (not shown in all studies), and lower rates of prescription issue (especially antibiotics), and more preventive activities occurring. Mean face-to-face consultation time is 8min for 10min appointments but only 9.2min for 15min appointments suggesting extra time may not be well used by doctors when booking interval exceeds 10min. Running late is stressful for doctors (and patients): it is easier keeping to time for 10min (rather than shorter) bookings. Other factors apart from time of year and social setting which increase (↑) or decrease (↓) consultation rates:

- List size (↑, ↓), and having personal lists (↓ by 7%).
- Not prescribing for minor ailments—see p 458 (↑↓)
- New patients (first year), and patients over 65yrs (↑).*
- If the GP is extrovert he recalls more, and his rate is higher than others (eg 6/yr vs 2/yr). GP age and sex also influences rates.
- High latitudes—within UK (↑). The South-East has lowest rates.¹
- Social deprivation (↑) and morbidity (↑). Rates fell in the 1960s–70s, perhaps because the burden of respiratory disease lessened.
- Preventive activities (↑). Non-attenders are now invited to clinics.

There is good evidence for the Howie hypothesis that average time of consultations is a valid and measurable marker of quality of care; see p 458–9.

*In females, the UK consultation rate at 10yrs is ~ 2.4 /yr; at 20 it is ~ 5 /yr, at 80 it is 7/yr. In males, the consultation rate at 10yrs is 2/yr; at 20 it is 1.7/yr; at 50yrs it is 2.7, at 70 it is 5.6 at 80 it is 6.7/yr. A GP with a list of 2000 with 300 patients >65yrs old will have provided 210 more consultations to this age group in 1998 than in 1992. S Rowlands 2002 *BJGP* 656

Phone consultations: is saving time or creating problems?

Telephone consultations and telephone triage (p 787) may seem a tempting way to reduce the need for precious appointment slots, and hence to move towards the UK government's 48-hour access targets. What is the evidence about this? First of all, during phone consultations, most non-verbal cues are missing. To explore this issue in a practical way, try consultations with friends or actors with whom you are sitting back-to-back. One way to improve these consultations is to do more 'explicit categorization', eg 'First I am going to find out more about how you are now, then I'll ask about your drugs, then I'll go over what I can do to help, then what to do if things get worse...'

Research shows that use of telephone consultations in place of same-day, face-to-face consultations does save time—but this time-saving is likely to be offset by higher re-consultation rates and less use of opportunistic health promotion.¹

1 B McKinstry 2002 *BJGP* 52 306

Medical certification, living wills, and other forms

Incapacity sick pay We despatch the metaphysical task of deciding who is well and who is sick, and for how long with amazing (but spurious) precision: appendicectomy \approx 2–3 weeks; open hysterectomy or CABG \approx 6 weeks; MI \approx 5 weeks; cholecystectomy \approx 2 weeks (5 weeks if open); laparoscopic inguinal hernia \leq 2 weeks (with driving; longer with older techniques).

www.dss.gov.uk/publications/dss/2001/medical/hot.htm

In England, form **SC1** can be used for self-certification for 7 days (got from surgeries, post offices, or benefit offices). For longer periods, GPs fill out **Med3** if seeing the patient to-day (or yesterday). If the patient has an illness that keeps them at home, don't feel you have to visit just for the sake of being able to say you've seen them for Med3—use form **Med5**; a patient can come in after a period of illness, and it can be back-dated. It can also be used without you seeing the patient if the patient has had a period in hospital (name the consultant). **Med6** can be used where there is a problem informing the employer of the exact nature of the illness. It can receive a vague diagnosis, and tells Social Services to send a separate form for the exact diagnosis. Social Services may also ask you to fill in a one-off form **Med4** (personal capacity) for further details of an illness.

Form **MatB1** gives a pregnant woman time off work once she is 14 weeks from the beginning of the week of her expected date of confinement. It may be signed by a GP or midwife.

Enduring power of attorney This legally passes authority over conducting one's financial affairs to a named person, who can then sign cheques, for example, for the other person. It holds good if, for example, the patient has a stroke or gets demented—provided the patient was fully aware at the time it was drawn up.

medline:8715552

Living wills Our practice keeps advance directives which limit the care a patient will accept after a mentally incapacitating illness such as a stroke. They have clauses such as 'after a stroke from which there is no prospect of recovery, I decline to be tube fed with nutrition, even if this hastens my death. I understand what tube feeding entails; it has been explained to me by my GP, Dr... on [date], and it is my considered and enduring wish that...' These documents have (some) legal force, and we must record on the patient's record that one is in existence. Copies should be distributed to the family and solicitor.

medline:12038510

The consultation

The consultation is the central act of medicine: all else derives from it. When medical errors occur, the reason usually lies in a failure of some process of the consultation. We must acquire flair for telling which part of which model is vital at any time, so that in busy surgeries with urgent visits mounting up, both the doctor and his or her patients can survive.

A medical model This is a way of describing what happens in a consultation in purely medical terms: History → examination → investigation → diagnosis → treatment → follow-up.

A patient-centred model¹ This comprises 8 stages:

The GP encourages the patient's contribution to the consultation.

The GP sets the patient's complaint in social and psychological contexts.

The GP explores and tests patient's ideas, concerns, expectations, and health beliefs. These beliefs are used to explain diagnosis & treatment.

The GP gets sufficient information for no serious condition to be missed.

Physical exam either addresses patient's concerns, or confirms or refutes hypotheses generated by the history, leading to shared working diagnosis.

The patient participates in the planning of treatment. Any prescription, referral, etc reflects best practice.

The GP aids *concordance* (formerly *compliance*, p 460) over therapeutic issues.

The GP establishes rapport with the patient, with appropriate follow-up.

Stott & Davis model²

Managing presenting problems	Modifying help-seeking behaviour
Managing continuing problems	Opportunistic health promotion

A hypothesis-testing model Information is collected and its validity is ascertained by generating and testing hypotheses.

Goal models (ie ends matter, not means) Aim to:

- Cure; comfort; calm; counsel; prevent; anticipate; explain.
- Enable the patient to put himself back in control of his life.
- Manipulate society to the patient's advantage.
- Facilitate change where change is what the patient desires.
- Increase patients' stature—by tapping the sources of richness in their lives, so freeing them from the shadow of insoluble problems.

The inner consultation (Roger Neighbour) model³

1 **Connecting** is establishing rapport.

2 **Summarizing** is the point at which the patient's reasons for attending, his hopes, feelings, concerns, and expectations have been well enough explored and acknowledged for the consultation to progress.

3 **Handing over** follows the doctor's assessment and diagnosis of the presenting problems and explained negotiated and agreed management plan.

4 **Safety netting** allows the doctor the security of knowing that she has prepared, or could prepare for, contingency plans to deal with an unexpected event and some departures from the intended management plan.

5 **Housekeeping** allows the GP to deal with any internal stresses and strains.

In consultations that are going wrong, ask yourself:

'Am I granting as much space to the patient's agenda as to mine?'

'Have I discovered his hopes and expectations—and his fears?'

'Am I negotiating openly with the patient over our clashing ideas?'

'What are my feelings, and how can they be used positively?'

Try saying: 'Things aren't going very well. Can we start again?'

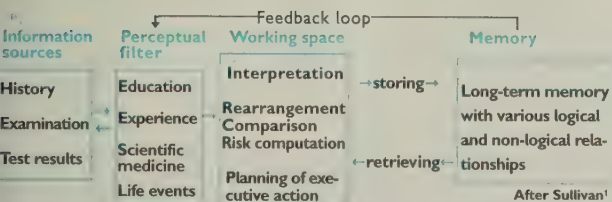
Unconscious consultation and the perceptual filter

What we want to examine here is that mental set and 'perceptual filter' which determines the outcome of consultations. When decision-analysts started work observing consultations they were amazed at the number of decisions per minute, and the wide range of possible outcomes, such as 'no action; review next week' or 'blue-light ambulance direct to tertiary referral centre' or 'refer to nurse for ECG' or 'prescribe x, y, and z, and stop Q in a week if the blood-level is such-and-such'. The average decision-analyst is disorientated by the sheer pace and apparent effortlessness of these decisions—so much so that doctors were often suspected of choosing plans almost randomly, until the idea of a 'perceptual filter' was developed.

Perceptual filters This is the internal architecture of our mind—unique to each doctor—into which we receive the patient's history. It comprises:

- Our unconscious mental set (*tired/uninterested to alert, engaged, responsive*).
- Our entire education, from school to last night's postgraduate lecture.
- The sum of all our encounters with patients. Ignore the fact that we can recall very few of these: this does not stop them influencing us strongly: does the rock recall each of those many, many waves which have sculpted it into extraordinary shapes, or which have entirely worn it away?
- Our past specific, personal experience with this particular patient.
- Our past specific, personal experience with the disease(s) in question.
- Non-personal *subjective* (eg 'endocarditis is the most dangerous and stealthy disease. . .') or *objective* ideas (eg evidence-based medicine, p 433).

The mind's working space (ram, not rom + memory, rom) The perceptual filter achieves nothing on its own. What is needed is: interpretation, rearrangement, comparison, and planning of executive action. • *The abilities of our mental working space are determined by the number of items of data that can be integrated into a decision.* There is evidence³ that this vital number is, for us, ~3 to 8 (this is the number of factors that can be considered *simultaneously*: like a juggler, you may have a hundred balls, but you can only keep 8 in the air at once). So, in trying to decide whether to start antidepressants, I might weigh the depression's severity (eg early waking with poor appetite and concentration), the patient's past omnivorous attitude to her pills (3 past overdoses), her expectations about treatment, the disadvantages of remaining depressed (eg child abuse), and knowledge that beating depression is a UK 'Health of the Nation' target. There may be just room enough in my 'RAM' to take into account evidence-based medicine on whether SSRIs (p 341) are likely to lead to deaths from overdose. The interesting experimental point here is that if we overload our 'RAM' in the consultation by recourse to a drawer full of guidelines, or unfiltered information—eg looking things up in an unstructured textbook, performance may decline.³



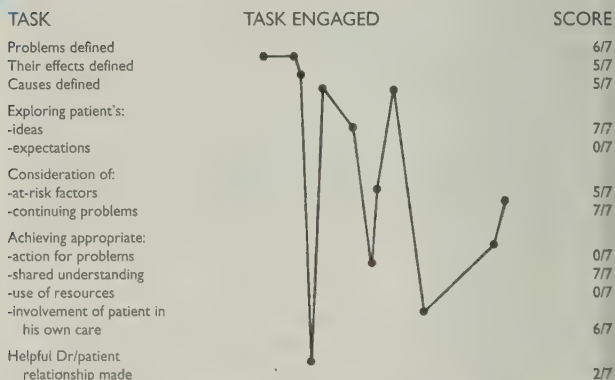
Consultation analysis

It is a sad fact that we lose some of our innate skills in communicating while at medical school. Consultation analysis aims to revive and extend this art, and we know it brings permanent improvement in those who participate. But do not conclude from this that research into the perceptible surface of behaviour can ever fully show us what is happening in the consultation.^{1,2}

Methods The first step is to gain the patient's consent. The method which gives the most information and the most scope for learning employs an observer/director sitting behind a 2-way mirror, who can pass verbal instructions to the doctor through an earphone which is worn unobtrusively. The activity is videotaped for later analysis. By directing the verbal and nonverbal behaviour, the observer can demonstrate the potential of a consultation in ways that the doctor may not have imagined possible. Other methods include simple videotaping, audiotaping, and joint consultations, in which the second doctor either participates in or observes the first doctor's consultations.

Consultation analysis is likely to be a somewhat threatening activity, so rules have been evolved to minimize this.³ For example, facts are discussed before opinions, the consulting doctor says what he did well, and then the group discusses what he did well. Then the consulting doctor says what he thinks he could have done better, and finally the group says what he could have done better. In practice, these constraints are occasionally stultifying, but it is better to be stultified than hurt.

Mapping the consultation and scoring its effectiveness In the consultation mapped below, the patient's inferior myocardial infarction (sudden chest pains on swallowing hot fluids) was mistaken by the doctor (JML) for indigestion, illustrating that there is no point in being a good communicator if you communicate the wrong message. It also shows how misleading it is to add the scores (50/84, but the patient nearly died).



1 J Skelton 1999 *Lancet* 353 108. This paper uses concordances of serial consultations to investigate such things as the play of power in doctor/patient relationships, and shows that the old schools of behaviour-based and meaning-based analysis need not be mutually antagonistic. 2 N Chomsky 1965 *Aspects of the Theory of Syntax*, Cambridge. 3 S Kurt 1998 *Teaching and Learning Communication Skills in Medicine*. Radcliffe Medical Press.

On the impossibility of being a good doctor

Here is a list of some of the things pundits tell us we should be doing when we meet patients^{1,2} (don't get depressed yet: we promise there *is* a solution to prevent any mounting feelings of inadequacy):

- 1 Listen to the history without interrupting or taking control of the agenda (how often are we guilty of indicating to the patient: 'don't talk to me when I am interrupting you')?
- 2 Examine the patient thoroughly (a nonsensical idea, or at least, so it would seem to the average patient with sciatica when you ask them to name the parts of a clock, or to pronounce 'The British Constitution').
- 3 Arrange cost-effective investigation (via a trip to the library if needed).
- 4 Formulate a differential diagnosis in social, psychological, and physical terms (a famous triad, no doubt, but why exclude spiritual, allegorical, materialistic, metaphysical, and poetic dimensions of the consultation?).
- 5 Explain the diagnosis to the patient in simple terms (then re-explain it all to relatives waiting anxiously outside, and then try re-explaining it to the computer in terms it understands—ie 5-digit Read codes).
- 6 Consider additional problems and risk-factors for promoting health.
- 7 List all the treatment options, and seek out relevant systematic reviews, guidelines, clinical trials, etc. (evidence-based bedside medicine).
- 8 Incorporate the patient's view on the balance of risks and benefits, harmonizing his view of priorities, with your own assessment of urgency.
- 9 Arrange follow-up and communicate with all of the healthcare team.
- 10 Arrange for the purchase of all necessary care, weighing up cost implications both for your other patients and the whole community, being prepared to be held accountable for your actions and inactions, and for the efficient use of resources—with recourse to justifications based on explicit criteria, and the principles of autonomy, non-maleficence, beneficence and distributive justice.

The alternative Look the patient in the eye. Look the disease in the eye, and then do your best.

The synthesis The alternative looks promising—even attractive, when compared to the ten impossibilities above. But note that the alternative only looks attractive because it is vague. 'Do your best' is not very helpful advice—and once we start unpacking this 'best' we start to get a list like the ten impossibilities. So... the answer is to rejoice in the fact that we are members of a profession where top marks go to the person who knows what *should* be done, not simply what *can* be done. In other words, we can rely on our own instinctive flair and judgment to decide which, if any, of the ten impossibilities we should address in any given consultation.³ What we cannot leave to our own instinct is the furnishing of our mind so that, should the need arise, we are *equally confident* in negotiating our way around *any* of the ten obstacles above.

Prevention

► In all disease the goal is prevention. This is not to say that preventive activities are without side-effects. Classification: preventing a disease (eg by vaccination) is *primary prevention*. Controlling disease in an early form (eg carcinoma *in situ*) is *secondary prevention*. Preventing complications in those already symptomatic is *tertiary prevention*.

Prevention through the human life cycle Pre-conception (p 94). Is she using folic acid supplements? Is she rubella immune? If not, vaccinate—ensure effective contraception for 3 months after vaccination. Is she diabetic? If so, optimize glycaemic control as early as possible (p 156).

Infancy Vaccination (p 208); safety lessons; developmental tests (p 288).

Adulthood Cigarette smoking trebles the risk above the rate for men who have never smoked. A systolic BP >148mmHg (40% of men) doubles the risk, and if the serum cholesterol is in the top fifth of the observed range, the risk trebles. Help to stop smoking (p 452), and treating marked arterial hypertension (*OHCM* p 126) and hyperlipidaemia (*OHCM* p 708) are the chief interventions. In America, intensive dietary advice is recommended for those in the top ¼ of the distribution of cholesterol values (this would include 50% of all British men), using drugs for those in the top 10th who do not respond to diet alone. The UK government recommends reductions in total fat and saturated fat to 35% and 15%, respectively, of total energy. GPs and practice nurses have a central rôle in preventing cardiac deaths, eg by identifying and reducing risks in young, first degree relatives of those who have had early coronary events (<50yrs), screening for hypertension, and encouraging less smoking. The fact that the optimum diet for reducing mortality has yet to be declared does not justify inaction with regard to the above preventative measures.

Postmenopausal women Education and self-examination.* Well-woman clinics. Mammography (using negligible radiation)—cancer pick-up rate: 5 per 1000 'healthy' women screened. Yearly 2-view mammograms in postmenopausal women might reduce mortality by 40%—but the price is the serious but needless alarm caused: there are ~10 false +ve mammograms for each true +ve result. The UK national health service offers 3-yearly single views to those between 55 and 64 years old (older women may be screened if they want, but they have to 'opt in').

Sexual health (p 432): Safer sex education starting in adolescence (teaching to use condoms need not increase rates of sexual activity); family planning (p 60–8), antenatal/prenatal care (p 94, eg folic acid), screening for cervical cancer (p 34), blood pressure, rubella serology.

Diet Antioxidants, eg vitamin E, C, carotenoids, flavonoids, and selenium (*OHCM* p 698) are found in diets rich in fruits, vegetables, grains, and nuts—and are thought to protect from various diseases (*OHCM* p 98: free radical action on LDL is central to atherogenesis), diabetes mellitus, cancer, and Alzheimer's (see p 352 for benefits of ginkgo biloba, extracted from the maidenhair tree).

P Bars 1998 E-BM 2 80

Life style 'Keep fit', pre-retirement lessons, bereavement counselling visits from the health visitor may help in preventing disease. HRT (p18) also has a central rôle. But the main aim is to adopt the measures outlined above to ensure that there is an old age in which to prevent disease.

Side-effects No intervention is without side-effects, and when carried out in large populations the problems may outweigh the benefits.

Smoking and prevention: p 452. Seat belts and prevention: p 781.

Barriers to prevention

Cynicism (See opposite.) When, if ever, we contemplate cataclysmic, but preventable, ill health in ourselves, we may either believe that 'It won't happen to me' or we deliberately dare fate to make it happen to us. To some people, over-zealous and sanctimonious-sounding hectoring from bodies such as the UK Health Education Authority creates barriers to prevention, inciting anger and rejection by those who resent their taxes being spent by some State Nanny who assumes that all her charges are 'backward 5-year-olds' who cannot be trusted to think for themselves.¹ So people are now proud to announce that '... I eat everything, as much butter and fried foods as I can get. ... I smoke 40–60 cigarettes a day. ... To eat cornflakes, you've got to have sugar on them, and lots of cream, otherwise there is no point in eating them. ... As long as you keep smoking cigarettes, and drink plenty of whisky, you'll go on for ever.'

Personal self-interest All of us at times are prone to promote our own destruction as keenly as we promote our own survival. Knowing that alcohol may bring about our own destruction gives the substance a certain appeal, when we are in certain frames of mind—particularly if we do not know the sordid details of what death by alcohol entails. It provides an alluring means of escape without entailing too headlong a rush into the seductive arms of death. Gambling and taking risks is all part of this ethos.

Logistical barriers A general practice needs to be highly organized to be in a state of perpetual readiness to answer questions like 'Who has not had their blood pressure checked for 5 years?' or 'Who has not turned up to their request to attend for screening?' or 'Who has stopped sending in for their repeat prescriptions for antihypertensives?' On a different front, providing a sequence of working fridges in the distribution of vaccines to rural tropical areas poses major logistic problems.

Political barriers It is not unknown for governments to back out silently of preventive obligations as if influenced by pressure groups who would lose if prevention were successful. Some countries are keener to buy tanks than vaccines.

Economic barriers If child benefits were available only to those children who had had whooping cough vaccine, much whooping cough would be prevented. This approach is not popular in the UK.

Financial barriers A practice may have to pay for extra staff to run an effective screening programme. Coronary artery bypass grafts may prevent some consequences of ischaemic heart disease, but they are too expensive to use on everybody whom they might benefit.

Time barriers As we rush out of morning surgery to attend the latest 'coronary' we use up energy which might have been spent on studying patients' notes in the evening to screen to prevent the next one. Changing from a crisis-led work pattern to strategic prevention is one way that practice nurses can lead the way. There is some evidence that they are particularly successful at the meticulous, repetitive tasks on which all good prevention depends.

Metaphysics at the bedside—our own world's worst patient

We often find ourselves sitting on beds trying to persuade wayward people to courses of preventive action which will clearly benefit them, usually at some distant time in the future. We think this very clarity should be enough to persuade the person to act. But, as we dismally stamp our feet on the bare boards of our impatience, we resign ourselves to the fact that action will not follow. Why is this so often the case?

The first person to know the answer to this question was the world's worst patient and our own greatest poet: great not because of his mastery of his world, but because, as the world used him, often cruelly, and as his London doctors gave him up as a hopeless addict, he took on all our petty confusions and made them human, compelling, and universal. Samuel Taylor Coleridge answers our question in this way:

'To love our future Self is almost as hard as to love our Neighbour—it is indeed only a difference of Space & Time. My Neighbour is my other Self, 'othered' by Space—my old age is to my youth and other Self, 'othered' by Time. . .'

See Coleridge ii p 225, R Holmes

By being consumed by the fires of his addictions this poor poet became the burning wick in the lamp we now use to illuminate our patients' frailties.

Screening

This entails systematic testing of a population, or a sub-group for signs of illness—which may be of established disease (pre-symptomatic, eg small breast cancers), or symptomatic (eg unreported hearing loss in the elderly).

Principles of screening for a disease (eg a disease that is asymptomatic)

- The condition screened for should be an important one.
- There should be an acceptable treatment for the disease.
- Diagnostic and treatment facilities should be available.
- A recognizable latent or early symptomatic stage is required.
- Opinions on who to treat as patients must be agreed.
- The test must be of *high discriminatory power* (below), *valid* (measuring what it purports to measure, not surrogate markers which might not correlate with reality) and be *reproducible*—with safety guaranteed.
- The examination must be acceptable to the patient.
- The untreated natural history of the disease must be known.
- A simple inexpensive test should be all that is required.
- Screening must be continuous (ie not a 'one-off' affair).

Summary: screening tests must be cost-effective.

Disadvantages of screening tests Before offering screening, there is a duty to quantify for patients their chance of being disadvantaged by it—from anxiety (may be devastating, eg while waiting for a false +ve result to be sorted out) and the effects of subsequent tests (eg bleeding after biopsy following an abnormal cervical smear), as well as the chances of benefit. We are all guilty of exaggerating benefits and avoiding discussion of controversial areas with patients.

www.txt.co.uk/full/cfm?1040

Comparing a test with

TEST RESULT	Subjects appear to have the condition	Patients with condition	Patients with- out condition
	Subjects appear not to have condition	True +ve (a)	False +ve (b)
		False -ve (c)	True -ve (d)

How reliably is the test +ve in the disease? $a/a+c$

How reliably is the test -ve in health? $d/d+b$

Screening for disease

Cervical smears (if >20yrs, p 34)

Mammography (after menopause)¹

Finding smokers (+quitting advice)

Looking for malignant hypertension
(lesser hypertension is problematic)

Opportunities for effective screening

Mental test score (dementia, p 352)

Urine tests (diabetes; kidney disease)

Antenatal procedures (p 95)

Elderly visiting to detect disease

Sigmoidoscopy for rectal cancer²

When is screening worth doing? If screening is to be done at all, it

makes economic sense to do it in general practice. In the UK ≥ 1 million people see GPs each weekday, providing great facilities for 'case-finding' (90% of patients consult over a 5-yr period). Provided the GP's records are adequate the remaining 10% may then be sent appointments to attend for special screening sessions—eg at the well-woman clinic (p 432). Private clinics do limited work, eg in screening for cervical cancer, but there is no evidence that their multiphasic biochemical analyses are effective procedures.

¹ C White 2002 *BMJ* i 933 ² D Ranstorf 2002 *Lancet* 359 1267

*There is evidence that some screening causes morbidity (mortality-awareness and hypochondriasis[†])—so why is screening promulgated? One reason is that it is easier for governments to be optimistic than to be rigorous.

Problems with screening

From a report on cervical screening:* 'By offering screening to 250,000 we have helped a few, harmed thousands, disappointed many, used £1.5m each year, and kept a few lawyers in work.' Typical problems are:

- 1 Those most at risk do not present for screening, thus increasing the gap between the healthy and the unhealthy—the *inverse care law*.
- 2 The 'worried well' overload services by seeking repeat screening.
- 3 Services for investigating those testing positive are inadequate.
- 4 Those who are false positives suffer stress while awaiting investigation, and remain anxious about their health despite reassurance.
- 5 A negative result may be regarded as a licence to take risks.
- 6 True positives, though treated, may begin to see themselves as of lower worth than hitherto.

► Remember: with some screening programmes of dubious value, *it may be healthier not to know*.

*www.txt.co.uk/full.cfm?1040

Primary care and shared-care clinics

Examples of mini-clinics which are conducted in general practice

- Well-woman/well-man clinic
- Elderly 'nonattending' patients
- Giving-up-smoking clinic (p 452)
- Joint outreach clinics with a consultant who shares care (eg orthopaedics)
- Antenatal clinic
- Hypertension clinic
- Citizen's advice clinic
- Diabetes clinic medline 11871208
- Asthma clinic

Advantages of mini-clinics

- Easy to keep to management protocol
- Check-lists prevent omissions
- Co-operation cards allow shared care
- Flow-charts to identify trends
- Help from specialist practice nurse
- Fewer referrals to outpatients (↓by 20%)
- Better co-operation with hospitals
- GPs can improve clinical skills medline:7475592
- Dialogue with on-site specialist does improve treatment and referrals medline:7475592
- Extra time needed
- Extra training needed
- Not holistic
- Not flexible
- Value often unproven
- Access to hospital technology↓
- Travelling time by consultants to outreach clinics is wasteful

Disadvantages

It is likely that starting a clinic is a good (but not the only) method of implementing the advantages above. The main disadvantage is the temptation for doctors to manage the disease, and to forget about the patient who suffers it: 'I've had enough, doctor. I want to kill myself.' 'Yes, well, your blood sugar is not too bad. Any spells of hypoglycaemia recently?'

Activities in well-woman clinics

- Cervical smear and breast examination
- Breast examination/mammography
- Pre-conception counselling (p 94)
- Antenatal and postnatal care
- Rubella and tetanus* vaccination
- Smoking and alcohol advice*
- Safer sex advice for HIV*
- Family planning/sterilization*
- Diet and weight*
- Blood pressure*
- Discussion of HRT issues

There is disagreement about whether this is desirable, and, if so, whether we can delegate this to nurses.

There is disagreement about whether this is desirable, and, if so, whether we can delegate this to nurses. Some (but not all) studies report that cancers in those having this protocol are detected earlier, with improved survival curves, compared with mammography alone. But UK DoH advice is against breast palpation in asymptomatic women, even if on the Pill or HRT. The DoH advises against delegation to nurses, but in some practices it may only be nurses whom women find acceptable, so validating nurse training is a key issue.

Well-men Women live longer, so why should they get all the prevention? (it might be argued). Nurses can do all the well-woman activities *starred above in well-man clinics. One such clinic yielded ≥25% obese; 14% with diastolic BP ≥100mmHg; 66% needing tetanus vaccination; and 29% needing smoking advice. See the OXCHECK study, p 439 and the NHS National Framework for ischaemic heart disease (OHCM p 75).

2001 meta-analysis
medline:11679383

Diabetic clinic Education and encouragement are the most important activities. Advantages over hospital clinics: patients see the same person each time; weekly appointments are possible during periods of difficult control; telephone advice is easily available. Mini-clinics are cheaper than outpatient clinics and outcome studies have 'proved' effectiveness. medline-11679387

Even young insulin-dependent diabetics can be managed wholly in primary care from presentation provided there is no overt ketoacidosis. There are dangers in adhering too closely to protocols (p 434). However, the vital test is retinal photography or dilating the pupil for fundoscopy (p 476 & p 508). Other vital areas are diet, exercise & smoking advice, BP & lipid control, and round-the-clock blood glucose monitoring, with checks on U&E and HbA1c.

May prevent hospital admission.

Evidence-based medicine (E-BM)

This is the conscientious and judicious use of current best evidence from clinical care research in the management of individual patients. The problem: 2,000,000 papers are published each year. Patients may benefit directly from a tiny fraction of these papers. How do we find them?

A partial solution 50 journals are scanned not by experts in neonatal nephrology or the left nostril, but by searchers trained to spot papers which have a direct message for practice, and meet predefined criteria of rigour (below). Summaries are then published in *Evidence-based Medicine*.

Questions used to evaluate papers 1 Are the results valid? (Randomized? Blinded? Were all patients accounted for who entered the trial? Was follow-up complete? Were the groups similar at the start? Were the groups treated equally, apart from the experimental intervention?) 2 What are the results? (How large was the treatment effect? How precise was the treatment effect?) 3 Will the results help my patients (cost-benefit sum).

Problems with the solution The concept of scientific rigour is opaque. What do we want? The science, the rigour, the truth, or what will be most useful to our patients? These may overlap, but they are not the same.

- Will the best be the enemy of the good? Are useful papers rejected due to some blemish? Answer: *all* evidence need appraising (often impossible!).
- By reformulating patients in terms of answerable questions, E-BM risks missing the point of the patient's consultation. He might simply want to express his fears, rather than be used as a substrate for an intellectual exercise.
- Is the standard the same for the evidence for *all* changes to our practice? For example, we might want to avoid prescribing drug X for constipation if there is the slimmest chance that it might cause colon cancer. There are many other drugs to choose from. We might require far more robust evidence than a remote chance to persuade us to do something rather counter-intuitive, such as giving heparin in DIC. How robust does the data need to be? There is no science to tell us the answer to this: we decide off the top of our head (albeit a wise head, we hope).
- EBM is a lucky dip if gathering *all* the evidence on a topic proves impossible.
- What about letters columns? It may be years before fatal flaws are aired.
- There is a danger that by always asking 'What is the evidence. . .' we will divert resources from hard-to-prove activities (which may be very valuable) to easy-to-prove services. An example might be physiotherapy for cerebral palsy. The unique personal attributes of the therapist may be as important as the objective regimen: she is impossible to quantify. It is a much easier management decision to transfer resources to some easy-to-quantify activity, eg neonatal screening for cystic fibrosis.
- Evidence-based medicine can never be always up to date. Reworking meta-analyses in the light of new trials takes time—if it is ever done at all.

Advantages of evidence-based medicine • It improves our reading habits. • It leads us to ask questions, and then to be sceptical of the answers. • As taxpayers, we should like it (wasteful practices can be abandoned). • Evidence-based medicine presupposes that we keep up to date, and makes it worthwhile to take trips around the perimeter of our knowledge. • Evidence-based medicine opens decision-making processes to patients.

Conclusion There is little doubt that, *where available*, EBM is better than what it is superseding. It may not have much impact, as gaining evidence is time-consuming and expensive. Despite these caveats, evidence-based medicine is one of the most exciting recent medical developments. Let's all join in by subscribing to ideals and its journal.

Protocols and guidelines

- ▶ Freedom from only doing ordained tasks is essential for mental health.
- ▶ Beware accepting a protocol without knowing if it will affect your sympathy and time to communicate. Is the protocol independently validated. What is its *objective* (cost-containment, conformity, or care-enhancement?).
- ▶ Reject any protocol which does not specify conflicts of interests: most protocols (87%) are written by people with financial links to drug companies.¹

Sympathy is a delicate flower which has often withered before the end of morning surgery. If a protocol says that you must do 10 things to Mrs James who happens to have diabetes, both of you may be irritated by item 5: the doctor is running out of time, and the patient is running out of goodwill. She is really worrying about her husband's dementia, having long-since stopped worrying about her own illnesses. She does not really mind being assailed by lights, forks, stix, and lancets, if this is the price that must be paid for a portion of her doctor's sympathy. But if she finds that this sympathy has withered, who knows what her feelings may be, and how she will view her doctor?

Guidelines are seen as friendly, if flexible² and allow for the frailties of clinical science as it meets bedside reality; they can also be interactive, if instituted in a computerized record during the consultation.² Protocols, particularly if they have been handed down from some supposedly higher authority, have a reputation for being strict, sinister, and stultifying instruments for thought-control. How well do these stereotypes stand up in practice? It is known that doctors working in highly regulated environments with strict protocols perform suboptimally.^{3,4,5} It is also worth noting that very few laws are flexible: those which are, are dangerous because they invite abuse. (All men are equal, but. . .). But what laws *can* do is to state when they don't apply. This doesn't necessarily make them mere guidelines. You could say, that patients have a right to be asked if they want to participate in a protocol, and if they do, that it should be done properly. But what if it is the child's birthday today, and he really does not want to have his finger pricked for a glucose test, but he is most willing to go along with all other aspects of a protocol? If you are flexible, the price may be ketoacidosis after the party. Herein lies the paradox of protocols. They are designed to remove the many indefensible inconsistencies found in clinical medicine, yet the protocols depend on the individual doctor's own flair and instinctive judgment to be applied in the best way.

Perhaps the best approach is to welcome the good protocols, and develop some sort of meta-protocol which should be answered whenever (or almost whenever) such protocols are not adhered to. Why did you not adhere to the protocol? Please tick the appropriate box. ☐ My own convenience. ☐ The patient's stated wish. ☐ The patient's stated wish after being given full information. ☐ The protocol is contraindicated in this case because my instinctive flair and judgement tells me so.

In conclusion, each protocol should come with a comment on the likely effect on the doctor's sympathy; how often, on average, one would expect the protocol not to be adhered to; and what resources and time will be needed. There should also be a statement about each element of the protocol, and just what benefit it is likely to confer—not on the ideal patient, but on the ones likely to be found in the population being offered the protocol. If a protocol does not come with these safety features, beware it, unless, that is, you have invented it yourself—for your own protocols will always have a special status, being infallible, ingenious, and innocuous.

The placebo effect

Placebo effects are very powerful and are important not just in research, but also in demonstrating to us how our demeanour may be just as important as the drugs we give. It is often said that if two obstetric units (for instance) do things quite differently (eg for or against artificial rupture of the membranes) then if one unit is vindicated by research, the other unit must be wrong. This need not follow—not just because the population served by the 'errant' unit may be different. The errant unit may believe it is a centre of excellence, and its staff may rupture membranes with joy in their hearts, knowing they are fulfilling their destiny as the best obstetric unit bar none. This feeling may communicate itself to labouring mothers, who, due to an interaction between communication, beliefs, cognition, and contractility, have their babies with few complications—so much so, that if the unit went over to the 'correct' method, their results might come to mirror their plummeting self-confidence. (This is an important reason for the failure of imposed protocols which look good on paper: see p 434.) Research in this area is very difficult to do, because you cannot easily control for 'joy in the heart'—but with care it can be systematically analysed: in a placebo-controlled study of antihypertensives the partners of the enthusiastic doctor broke the code, and told him that his experimental treatment appeared similar to existing treatments without telling him who was having the active drug, and who was having the placebo. ▶ From this point, there was an immediate, marked increase in BP in both groups, although the difference between the drug and the placebo was maintained.

medline:7880221 & 7802773

▶ Our beliefs and our behaviour sometimes matter more than our drugs.

Speculative mechanisms *Psychological* Expectancy, anxiety-reduction and Pavlovian conditioning may operate. *Psychosomatic linkages*. Endorphins, catecholamines, cortisol, and psychoneuroimmunology. medline:10923577
NB: It is *not* the case that there is a 'placebo-responsive' personality.

Don't assume we should give placebos to all our patients, and be enthusiastic about their likely effects. Most of this book is founded on the precept that we should be honest and straightforward with our patients. Nevertheless, it may often be unwise to share too many of our doubts with our patients, as demonstrated by the GP who randomly assigned his consulting style in those with apparently minor illness to a 'positive encounter' or a 'negative encounter'. In the former, patients were given a diagnosis and told they would be better in a few days, the latter group were told that it was not certain what the matter was. Just over 1/3 of the patients having negative encounters got better in 2 weeks, but over 2/3 of the positive group did so.

Nocebo effects Pharmacists often 'leaflet' customers with long lists of their drugs' side-effects, which, like Voodoo death, may become self-fulfilling prophesies. When one of the authors (JML) gives diamorphine iv to patients having a myocardial infarction, he says in a loud voice 'Your pain will end now' (maximizes placebo effect)—and, in a very quiet voice to the spouse, 'Get a bowl: he may be sick.'—to avoid the reverse 'nocebo' effect. We tend not to tell patients about side-effects such as impotence, justifying this dishonesty on the grounds that in such a sensitive area, the mere mention of impotence will bring it about. Central to this debate is the question of how paternalistic we should be. We do not know the answer, but simply recommend getting to know individual patients well, and having a flexible approach: in doing so we may occasionally strike the right balance.

Bedside manners in a multicultural world

How to avoid offending Eastern manners • Avoid prolonged eye contact and loud speech—which can indicate lack of respect.

- Control your gesticulations (the 'thumbs up' sign, for example, may be considered very rude).
- Do not expect an Afro-Asian to answer intimate questions, without first explaining about confidentiality.
- A psychiatric referral may destroy eligibility for marriage.

Hindu names • First names are often male and female, but middle names always denote sex. • First (eg Lalita) and middle (eg Devi) names are often written together (Lalitadevi). • When asked his name a Hindu is likely to give only his first 2 names, withholding his family name, to be polite. This can cause great confusion with receptionists, and duplication of notes.

Sikh names Few Sikhs have a family name. Singh and Kaur indicate only sex and religion, so that extra identification is needed on the notes (use the father's or husband's name also).

Muslim names Sometimes the last name is not a family name, and the first name is not the personal name. There may be no shared family name. The first name is often religious (eg Mohammed). Common female second names are Banu, Begum, Bi, Bibi, and Sultana. They are of as little help in identification as the title Ms is in UK names, but because they are given as second names they are often used for filing purposes (so other family members should be entered as well, to give extra identification).

Muslims and death (Muslims are the largest non-Christian UK sect).

- Religious observance requires prompt burial, not cremation. Washing and shrouding is an important ceremony done by elders of the same sex.
- The dead body is sacred, and never abandoned by relatives; so it is rare for undertakers to be involved. The body may be moved in a family van.
- Bereavement lasts for 3 days (prayers in the home are said almost continuously). Wives stay at home for up to 4½ months after death of a spouse.
- Shoes are to be taken off before entering the house of the dead.
- Transplantation may be fine for Arab, but not Indian Muslims (the latter may allow corneas; bone marrow does not count as a transplant).

How to avoid offending Western manners This is not just a question of 'please', 'thank you', 'after you', and 'I'd love to. . .' rather than 'I don't mind if I do. . .' with feigned enthusiasm—Westerners are notoriously sensitive to gaze and mutual gaze: not enough, and they think you are shifty; too much, and you are making unwelcome advances; the same goes with interpersonal distance: if you get too close, invasion is threatened.

Clinical phenomena • Centile charts are different (p 185).

- Bilateral cervical and inguinal lymphadenopathy may be normal in Asian and African children (or it may indicate TB).
- Genetic diseases may be common, eg sickle-cell anaemia in Africans and West Indians; haemoglobin E disease in Chinese.
- Unusual malignancies (oesophagus in Japan, tongue from betel nuts).
- The cow is sacred to Hindus—so do not offer beef insulin (or pork insulin to those of Jewish or Islamic religions).
- Some Asians will consider measles to be a curse from the Goddess Mata and will opt for an offering in the temple, rather than a visit to the GP.
- Some Muslims will avoid all alcohol (so do not offer tonics).
- Taking drugs may be a problem for Muslims during Ramadan.

Language, interpretation, and health

Dislocation, globalization, migration, refugee numbers, and war conspire to separate doctors and patients linguistically. 90 million people now live outside their country of birth. Heterogeneous populations need heterogeneous methods of health-orientated communication—in many languages. All too often interpreters, if available, are only on hand during office hours. Friends and relatives may be available, but confidentiality issues are important. The interpreter's own cultural prejudices may distort your questions, and even make them vacuous.

medline:8520310

When a child interprets for his mother's or father's mortal illness, many other unknowable issues are raised.

medline:9582130

Alternatives include telephone interpretation (eg with a hands-off conference telephone). Although nonverbal signs are lost, there may be advantages as some patients may say things to a telephone which they would be unhappy to say face-to-face.

medline:9485622

Whenever needed, ask for these services from Health Authorities/Primary Care Trusts—and then the services are more likely to be in place, and the wheels well-oiled when the real emergency arises.

The Pro and Con points for interviewing an interpreter

Address patient in the second person

Talk directly to your patient

Keep control of the consultation

Pause often, looking for nonverbal clues which may mean misunderstanding

Appear attentive when patient responds

Check your patient's understanding

Make full use of written material

www.bmj.com/cgi/content/full/311/7004/555

Health education

What is education?—four incomplete answers

- Education is the system used for passing down, from one generation to the next, society's values, attitudes, and culture. Thus are crime, duplicity and double standards (and, on a good day, idealism) perpetuated in a kind of cultural inheritance.
- Education is an activity carried out on ignoramuses by people who think they know better.
- Education is about changing people. It usually ends up implying 'change your ways . . . or else.' The most extreme form of education is prison.*
- Education performed on one mind by another, under duress, is indoctrination. Indoctrination has its uses. Its value is measured by its propensity to encourage self-education, through, for example, travel, reading, or dialogue. Self-education is the food of the mind: the procedure by which we can touch the great minds of the dead and know we are not alone in all our confusions and questionings. By standing on their shoulders we can find a new view of our world—sometimes, even, new worlds to view.

Health education messages To be effective, they must be specific and direct. Eg in getting people to sign on for help for drinking problems, it is of little use saying 'If you don't stop drinking you'll get these diseases. . .' (only ~25% respond); saying 'Signing on is good for you because of these benefits. . .' (~50% respond); saying 'If you don't sign here, you've had it. . .' brings the biggest response. A certain amount of 'fear' in the message is not bad: in enlisting patients for a tetanus vaccine a 'low fear' message gets a 30% response, while a more fear gets a 60%. Optimum messages must be very specific about dates, times, and places of help. Too high a level of fear is counter-productive. A gruesome film about the worst effects of caries produces petrified immobility, not self-help or trips to dentists.

The messenger Peers may be better than authority-figures (eg in stop smoking messages). A message about breast feeding will come best from a mother. However, if the issues are not well understood, authority may be helpful (the *BMJ* is more effective than *Woman's Own* in suggesting to mothers that a new formulation of aspirin should not be taken).

Changing attitudes The following paradigm holds sway: **knowledge→attitudes→intentions→behaviour**. As Chinese thought reformers knew so well, attitude changes depend on a high level of emotional involvement. In questions of belief, as in so many other questions, emotion trumps reason '*...people don't demand that a thing be reasonable if their emotions are touched. Lovers aren't reasonable, are they? . . .*'** Only resort to applying reason to attitudes if emotions are too hot to handle. NB: the arrows in the model above may be reversed: if our behaviour is inconsistent with our ideas (cognitive dissonance) it is often our ideas, not our behaviour which change.

Health education officers are likely to have a nursing, teaching, or health visiting background. They may have a postgraduate diploma in health education. There are ~300 in England and Wales. Teams comprise a technician and a graphic artist, as well as clerical staff. One rôle is to give information and Health Education Authority leaflets. They also liaise with health visitors and primary care trusts, as well as engaging in planning and research.

Examples of health education at work ● Education about 'safer sex' and the prevention of AIDS. ● Leaflets and tape/slide programmes can (slightly) increase knowledge of breast self-examination (which is associated with smaller tumours and less spread in those presenting with breast cancer). ● Radio dramas with health issues reaching millions, eg *Soul City* in South Africa.

Health promotion by nurses. Nurses are the experts in this field—but even they are not very effective in reducing coronary risk. In the community-based OXCHECK randomized trial ($N=6124$, aged 35–64) serum cholesterol fell by only 0.08–0.2mmol/L—and there was no significant difference in rates of giving up smoking, or in body mass index. Systolic (and diastolic) BP fell by ~2.5% in the intervention group receiving dietary and lifestyle advice. Blanket health promotion may not be a complete waste of resources, but it is certainly expensive for rather limited gains. Similar results have been obtained by the *Family Heart Study Group*. Depending on the assumed duration of risk reduction, the programme cost per discounted life year gained ranges from £34,800 if interventional benefits last for 1 year, compared with £1500 for 20 years duration. Corresponding OXCHECK figures are £29,300 and £900. These figures exclude broader longterm cost effects other than coronary mortality. medline:8634618

The conclusion may be that energies are best spent on those with highest risk as determined in routine consultations by a few ‘simple questions’ about smoking, family history, etc. One trouble is that these questions are not always innocuous. It is not necessarily a good thing to bring up ‘strokes and heart attacks in the family’ in, for example, consultations about tension headaches. OXCHECK is not the last word—and there is some evidence that if lipid-lowering drugs were used very much more extensively, cholesterol (and cardiac events) could fall by 30%. <http://bmj.com/cgi/content/full/310/6987/1099>

Novel ways of delivering health education messages

• For those who have difficulty in accessing health services, eg those in rural areas, videoconferencing and the internet may be a good way forward. Sustainability of these programmes depends upon the following issues: cost, delivery style, and availability of appropriate technology and patient-friendly internet sites. medline:11747664

• Traditionally, health education has been given by experts, partly because they can answer questions authoritatively. But this authority is itself a problem. Risk-takers are unlikely to listen to the prim and proper. So peer-education has been developed as a tool to reach certain groups, and evidence suggests that this is a promising way forward. medline:11728893

*P Theroux *Down the Yangtze*, Penguin ISBN 0-14-600032-3, page 35–6

**Graham Greene 1951 *The End Of the Affair*, page 115, Heinemann

Its themes are social & medical: child care, empowerment of women, & HIV (Garth Topham); evaluation shows a huge health impact, see www.cominit.com/idcc2001/sld-1765.html

Patient groups

Parent and baby groups These are best set up in the first weeks after the birth of four or five babies. The health visitor encourages the group to form. A doctor may attend the group—regularly to start with, then less often as the group becomes self-sufficient. After a year or two a large practice will have a number of groups running. One aim is to increase motivation (through discussion) to enhance the uptake of health education and preventative medicine. Another aim is to ease the stresses involved in becoming a responsible parent by providing a social support network. A mother, noting for the first time her beautiful baby's ability to hate, to destroy, and to hurt, may find it a relief to know that other babies are much the same.

Patient participation groups Working with your patients is as important as working for them. The health care team meets with patients' representatives to discuss some of the following:

- Dealing with complaints (less adversarial than with formal methods).
- Harmonizing the 'consumer's' and the 'provider's' aims.
- Feedback to aid planning, implementation, and evaluation of services.
- Identifying unmet needs (eg among the isolated elderly).
- Improving links between the practice and other helpers.
- Health promotion in the light of local beliefs (p 416).
- Pressurizing government institutions over inadequate services.

Owing to lack of interest, or to there being no clear leader or task, up to 25% of groups have closed. The complaint that participation mechanisms lead to tokenism (ie the democratic ideal has been exercised, but what has been created is just a platform for validating the *status quo*) does not turn out to be the case when a patient group has power over funds which it has raised. Here our experience is that analysis may be penetrating and decision swift, in a way which makes even the best-run health authorities/Trusts look pedestrian.

The patients' association This group represents and furthers the interests of patients by giving assistance, advice, and information. It aims to promote understanding between patients and the medical world. Publications: Patient Voice and a directory of self-help organizations. See also the Contact-a-Family Directory.

www.cafamily.org.uk/search.html

Self-help organizations Many thousands of these groups have been set up all over the world for sufferers of specific rare or common diseases. They offer information, companionship, comfort, and a life-line to patients and their families, eg for sharing techniques and self-remedies. A danger is that they share nightmares as well, for example, unnecessarily graphic descriptions of their children dying of cystic fibrosis may be spread, causing unneeded despondency. They raise funds for research, providing a 'welcome alternative to the expensive services of professionals'. Full directories exist (see above).

Tell someone close to the cause of your complaint such as a doctor, nurse, receptionist, or practice manager as soon as possible (within 6 months of the incident, or within 6 months of discovering that you have a problem). It is often possible to sort out the problem at once. This is called Local Resolution.* You can telephone or write to the practice complaints officer or to the individual concerned. However, if you would prefer to talk to someone who is not involved in your care, you can contact one of the NHS Trust Complaints Officers.

Complain to your Primary Care Organization or your own doctor. If your complaint is about , contact the Complaints Officer of your local Primary Care Organization.

Within 2 working days is the aim. For written complaints and cases where more detailed investigation is required, your complaint will be acknowledged in writing within 3 working days and the aim is to respond fully in 20 working days.

You may request an Independent Review. You must ask for this within 28 calendar days from the date of the letter responding to your complaint. You will be asked to write a letter to The Convener of the NHS Trust responsible for the service saying why you are still dissatisfied. The convener is a specially trained member of the NHS Trust and, having taken advice, will decide whether to refer the complaint back for further Local Resolution, possibly suggesting Conciliation—or to set up an Independent Review panel to consider the complaint; (Complaints Officers can explain to you how these processes work). You will be advised of the Convener's decision within 20 working days of your request (10 working days if about GPs, Chemists, or Opticians). www.lothianhealth.scot.nhs.uk/rights_complaints/making_complaint/

Should an Independent Review Panel be set up, this will consist of the Convener, an Independent Lay Chairperson, and one other person nominated by the NHS Trust. The Panel will investigate the complaint and talk to everyone involved, seeking the specialist advice it needs. You will then be sent a report, including the Panel's conclusions.

You may request an investigation by the Health Service Commissioner (ombudsman) who is independent of the NHS and the Government. NB: the General Medical Council can be involved with a complaint whether or not it relates to an NHS patient.

Complaint systems are only a part of clinical governance, a system, which links continuing professional development, multidisciplinary learning, audit, risk management, and critical incident reporting. It is a 'framework through which the NHS is accountable for continuously improving the quality and safeguarding high standards of care by creating an environment in which excellence will flourish.' The sequence is: defining quality→assuring accountability→improving quality.

medline:11700383 & www.txt.co.uk/full.cfm?1016

It is one mechanism by which the Commission for Health Improvement (CHI) operates. Its areas of priority are partly set by the National Institute of Clinical Excellence (NICE) and health improvement programmes (HIMP).

*It is the practice manager's duty to log each complaint with its outcome, to acknowledge written complaints within 3 days, and to send a copy of the Practice's Complaints Procedure leaflet. She ensures that the internal investigation takes ≤10 days (if longer, she must give reasons). She will take advice from the medical defence organization.

Dying at home

'Don't touch me! Don't question me! Don't speak to me! Stay with me!'*

The UK death rate is ~12/1000/yr, or ~30 deaths/GP/yr. 65% die in hospital, 10% die in hospices, public places, or on the street, and 25% die at home. Of these deaths at home over half will be sudden. In the remainder, the GP has a central rôle to play in enabling the patient to die a dignified death in the way that he or she chooses. Pain relief and symptom control are the central pre-occupations of death in hospices, where death has already been somewhat medicalized, but in those who choose to die at home there often runs a fierce streak of independence, so that their main aim is to carry on with the activities of normal living—come what may. This may cause distress to relatives who feel that the dying person is putting up with unnecessary pain. An open discussion is often helpful in harmonizing the family's aims. The important step is to find out what a patient wants, and then to enable him to do it, being aware that his aims may change over time. The next step is to find out about his hopes and fears and how they interact with those of the family.

Pain ▶ See OHCM p 426: *diagnose each pain*. Bone pain will need opiates, ± NSAIDs, eg naproxen 250mg/8h po pc or prednisolone 5mg/8h po; pain from constipation responds to laxatives or enemas. Document each pain's response to treatment. Morphine (oral) is given eg every 4h. Strength examples (/5ml): 10mg, 30mg, 100mg. Once the daily dose which controls pain is known, consider giving the same 24h dose as slow release tablets every 12h (eg MST® 5, 10, 15, 30, 60, 100, or 200mg). Take the first MST tablet 4h after the last dose of morphine solution. (Starting dose of MST in children: 200–800µg/kg/12h po.) If pain breaks through, give extra morphine solution, and ↑MST at the next dose (don't give more than twice daily to avoid pharmacodynamic confusion). If the patient cannot swallow, use diamorphine sc, at ½ the daily oral morphine dose, given eg divided into 4-hourly injections or via a pump. Laxatives ± antiemetics will be needed.

Help •Family •Community nurse •Hospice •Friends/neighbours
•Night nurses •GP/health visitor •Pain clinic •CRUSE (selfhelp group)

Bereavement is the process of adapting to a loss, which is causing sorrow.

Mourning is the active evocation of that loss, which may take a formal, ritualistic, and dignified style, or be personal and idiosyncratic. **Lamentation** is the vocal outpouring of grief at its cusp, with wailing, lachrymation, or verbal expressions uttered from the depths of despair. **Grief** has components of all these, but its defining feature is the sense of irreversible personal loss. ▶ *There is in this world in which everything wears out, everything perishes, one thing that crumbles into dust, that destroys itself still more completely, leaving behind still fewer traces of itself than Beauty: namely Grief.***

The bereavement process Numbness → denial → yearning → depression → guilt/aggression → reintegration. This process may take years. The process may become pathological if major depression (p 336) is precipitated. It is often tempting to try and 'do something' by giving psychotropics, but it is known that most bereaved people do not want this, and there is no evidence that drugs reduce problems. Empathy and helping the patient to shed tears is probably the most valuable approach, especially when losses are covert or hard to acknowledge. Counselling after bereavement is effective. medline:9582150 After bereavement, risk of death in spouses rises in the 1st 6 months (men) or in the 2nd year (women). Men and younger bereaved are at greatest risk. It is not known whether this is due to shared unfavourable environments or to psychological causes (eg mediated by the immune system). The main causes of death are vascular, cancer, accidents, and suicide. medline:3094678

- Distancing tactics: 'Everyone feels upset when there is bad news, but you'll soon get used to it.'
- False reassurance: 'I am sure you will feel better; we have good anti-emetics these days.'
- Selective attention: 'What is going to happen to me? I'm beginning to think I'm not going to get better this time. The pain in my hip is getting worse.' Doctor: 'Tell me more about your hip.'

Breaking bad news Bad news is any information that drastically alters a patient's view of their future for the worse. Patients have a right to such knowledge, but not a duty to receive it, so negotiation is needed to agree the type of information and the amount they want to hear at any moment. The advantage of patient's being aware of bad news are:

- It helps maintain trust in those caring for them
- To reduce uncertainty
- To prevent inappropriate hope
- To allow appropriate adjustment
- To prevent a conspiracy of silence

The central activities in the breaking of bad news are:

- 1 Preparation—choose a quiet place where you will not be disturbed.
- 2 Find out what the patient already knows or surmises.
- 3 Find out how much the person wants to know. You can be surprisingly direct about this. 'Are you the sort of person who, if anything were amiss, would want to know all the details?'
- 4 Fire a warning shot. 'I am afraid I have bad news.'
- 5 Allow denial.
- 6 Explain (if requested). Share information about diagnosis, treatments, prognosis, and specifically listing supporting people (eg nurses) and institutions (eg hospices). Try asking 'Is there anything else you want me to explain?'
- 7 Listen to concerns.
- 8 Recognize and encourage ventilation of the patient's feelings.
- 9 Summarize and make a plan.
- 10 Offer availability. The most important thing here is to leave the patient with the strong impression that, come what may, you are with him or her *whatever*, and that this unwritten contract will not be broken.

*Estragon in http://samuel-beckett.net/Waiting_for_Godot_Part2.html

**M Proust 1925 *Albertine Disparue*. NB: Proust's view is clear enough for grief surviving down generations—which it does not. But what of one person's grief? Surely this can last a lifetime, and Proust is wrong? No: we must assume that Proust had in mind an image from Gérard de Nerval (whom he rated very highly) who describes great grief as being carved into us, not like lettering on stone, which may be worn away by time, but like those initials we carve into the bark of a living tree. As the tree grows, the lettering sinks ever deeper, so that it may, apparently, to the casual observer, disappear, but really it has been embodied, internalized, and taken up into the inner life of the organism and distributed even to its fingertips (see Richard Holmes' on Gérard de Nerval in *Footsteps* 1996 HarperCollins, p 222).

Useful agents in terminal care*

Remember that pain has physical, emotional, and spiritual components: all aspects need to be addressed. Diagnose and monitor each pain separately.

Diamorphine po: 2mg=3mg morphine. sc: 1mg diamorphine=1.5mg morphine. Sustained release morphine is available, eg MST-30® (30–100mg/12h po) or MXL (lasts 24h). or can be used

when dysphagia or vomiting make oral drugs useless, eg oxycodone 30mg suppositories (eg 30mg/8h, ≈30mg morphine). If crystallization occurs with drugs mixed in syringe drivers, either increase the dilution, or use 2 syringe drivers.

are also useful, eg fentanyl (Durogesic® patches last 72h: typical starting dose for someone needing the equivalent of 90mg morphine/day would be one Durogesic-25® patch (the 25 means 25µg fentanyl/h; 50–135–224mg MST, 75–225–314mg MST and 100–315–404mg MST patches are also available). Use non-irritated, non-irradiated, non-hairy skin on trunk or upper arm; remove after 72h and replace by another patch at a different site.

may especially help (ibuprofen 400mg/8h po or diclofenac + misoprostol, as Arthrotec®, 1 tablet/12h po.) Pamidronate may also relieve pain from bony secondaries (OHCM p 698).

Amitriptyline 25–50mg po at night ± clonazepam 0.5–1mg/24h po nocte, increased slowly to 1–2mg/8h. —Nerve blocks are useful.

Low residue diets.

Co-danthramer capsules or liquid ± bisacodyl 5–10mg nocte.

eg associated with gastric carcinoma—H₂ antagonists (eg cimetidine 400mg/12h po or proton pump inhibitors (omeprazole).

in jaundice—Cholestyramine 4g/6h po (1h after other drugs).

or Buprenorphine sublingual 0.2–0.4mg/8h.

Not a pure agonist. 'Ceiling' effects negate dose increases.

Foul rectal discharges—Betadine vaginal gel®.

Cyclizine 50–150mg/4–8h po, pr, im, sc. Prochlorperazine suppositories: 25mg/12h. Haloperidol (p 360) 0.5–2mg po.

try hyoscine hydrobromide 0.4–0.8mg sc/8h or 0.3mg sublingual. Octreotide, max 600µg/24h via a syringe driver may remove the need for palliative surgery, ivs and NGTs.

Metoclopramide 10mg/8h po or sc. If this fails, try domperidone 60mg/8h rectally.

Thoracocentesis (bleomycin pleurodesis).

Air hunger: Chlorpromazine (eg 12.5mg iv).

Hyoscine (as above) 0.4–0.8mg/8h sc or 0.3mg sublingual.

(hypoxic)—Table fans ± supplemental humidified oxygen.

Haemoptysis: Diamorphine, above, iv if massive.

Intercostal nerve blocks may bring lasting relief.

often causes distressing symptoms. Try spironolactone 100mg/12h po + bumetanide 1mg/24h po.

Betadine vaginal gel®.

—Alum irrigation (1%) by catheter (in hospital).

may be cleaned by 6% hydrogen peroxide, chewing pineapple chunks to release proteolytic enzymes, sucking on ice, or butter.

—Steroids: dexamethasone is most useful: give 8mg iv stat.

Tabls are 2mg (≈15mg prednisolone) NB: dexamethasone given at night can prevent sleep.

Psychological symptoms

—Try diazepam 10mg suppositories (eg 10mg/8h), or haloperidol (p 360) 0.5–2mg po (may help nightmares, hallucinations, and vomiting too). Midazolam may be used in syringe drivers (eg 5–10mg/24h)—or levomepromazine (Nozinan®) 12.5–50mg im stat or 25–200mg/24h sc via a syringe driver.

—Steroids; most useful is dexamethasone, eg 4mg/12–24h po to stimulate appetite, reduce icp, and in some patients induce a satisfactory sense of euphoria.

UK patients and relatives may get much support via (Brit Assⁿ of Cancer United Patients, 3 Bath Place, Rivington St, London, EC2A 3JR, tel. 020 7613 2121). In the UK, have a vital rôle in fast-tracking applications from financial help in the form of the , and the , once the prognosis is <6 months. See p 462–3.

Sources: G Hanks 1996 *BMJ* **i** 823–26; *BNF* 2002 *Prescribing in Palliative Care* (ch 1); D Doyle 1994 *Oxford Textbook of Palliative Medicine*; P Kay 1994 *4–2 of Hospice and Palliative Medicine*, *MJ Baines Annals Acad Med* **23** 178–82 **Acknowledgement:** This page owes much to Dr Tim Wiggan.

Records and computers

- ▶ We cannot make ourselves better people by using a system or a machine: reflection, dialogue, and action are more likely routes to self-improvement.
- ▶ It is the care and interest a doctor takes in his records that matters, not the details of the particular system employed.

Problem-oriented records—*eg* 'breathlessness' (not 'bronchitis'—or 'Down's syndrome', which may be unproblematic to the individual, but a problem to us 'normals', who do not like disability: so often such a patient's *real* problem is that we have a problem).

How the patient and/or carer sees the problems.

Physical examination and results of tests (not always objective!).

Social, psychological, and physical interpretation.

Do the following tests. . . or 'Wait on events'; treatment: *eg* 'Start psychotherapy' and explanation—note what the patient has been told.

Computers Advantages include:

- Faster communication with laboratory and hospital.
- Better and *more* preventive care, not just in theory, but in practice (*eg* 8–18% more vaccinations in consultations with a computer on the desk, more BP measurements, with a ~5mmHg fall in BP in those with hypertension; the price is longer consultations—by 48–90sec).
- Audit: *eg* computerized record of everyone's BP and vaccinations.
- Prescribing: computers save money (*eg* 8%) by making generic prescribing easy. They are more likely to be associated with complete records (95% vs 42%). The electronic *BNF* allows simultaneous viewing of various sections, and 'hot-links' to drug interactions. Computer-assisted prescribing linked to Read codes (*eg* *PRODIGY*) can aid prescribing, and aims to improve quality by automatically linking prescribing to practice-modifiable guidelines.²
- Can send letters to patients, *eg* in the light of new drug information.
- Integration of data from multiple sources—*eg* drug interactions.
- Quick exploration of hypotheses. (Do I see more depression in winter?)
- Assessing eligibility for social security benefits.
- Rule-based systems/neural networks—is this ECG/smear abnormal?
- Keeping up-to-date, decision support and evidence-based medicine, *eg* the Internet,* and the *Oxford Handbook of Clinical Rarities*.^{3,4} the latter uses this text, *OHCM*, and the *Oxford Handbook of Clinical Rarities*.³ This database is regularly updated and contains >25,000 key-worded facts connected by an intelligent index, linked to the medical record via Read codes to enable explanation of apparently unrelated or obscure signs, symptoms and results, *eg* chest pain, depression, MCV↑ and melaena are explained by alcoholism, with 'postulates' of related trauma (fractured ribs) or cardiomyopathy—both known associations of alcoholism—to account for the chest pain.
- Electronic health records are a goal, the basics of which are easy to see (communicating prescriptions, appointments, lab and clinic data seamlessly between hospitals, GPs, pharmacists and patients—*eg* in the UK NHS via the NHS net). Implementation on a large scale is proving difficult, and it's easy to criticize the NHS for delays—but note that the UK is the only country to have an overarching information-technology strategy agreed.
- Paperless practices become possible; no lost notes; more space; less staff time spent filing—but more dependency on having skilled staff in place.

* F Sullivan *E-BM* 196 2 | Purves 1998 *JRCGP* 48 1552; M Johnston 1994 *Arch Int Med* 120 135

3 Mentor; EMIS, tel. +44 (0)1329 828533/1132 582454/ 01132 591122 www.mentor-update.com

4 *BMJ* 1993 ii 679 **Eg* Global health www.pitt.edu/HOME/GHNet/GHNet.html

Medline

12 million abstracts & full texts free at www.ncbi.nlm.nih.gov/pubmed

We all need to know about Medline—it's a skill as basic as taking blood—but easier. The **lucky-dip method** is good, but indiscriminating: just enter whatever you want in the search box, and press the 'go' button. NB: if this is **all** you want, and you require **only** full text, the best site is <http://pubmed.ncbi.nlm.nih.gov/>.

This page aims to help if you want to be sure you are not missing something, (and want to cut down irrelevant hits). If Medline is new to you, find a computer, and an internet-enabled friend to sit near you while trying out the searches below in **BLUE CAPTIONS**. When you click the 'go' button, the system tries to match your search to a nomenclature of **medical subheadings (MeSH terms)**. Suppose you're interested in alternative therapies for angina: type **ANGINA AND THERAPY** (use capitals; ► check your spelling!); click 'go', then the 'details' button to show that this is mapped to (('angina pectoris'[MeSH Terms] OR angina[Text Word]) AND (('therapy'[Subheading] OR 'therapeutics'[MeSH Terms]) OR therapy[Text Word])). There are >23,000 hits. Adding **AND RANDOMIZED** to the search phrase (the star gets *randomized, randomised*, etc) narrows these to <3000; adding **ALTERNATIVE** before the word **THERAPY** gives <40 hits.

Square brackets The contents of these limit or expand the search. Mostly, let Medline do this for you, as in the above example. But there are some square brackets it's useful to add to the search yourself. For example, **ANGINA AND BMJ** gives (('angina pectoris'[MeSH Terms] OR angina[Text Word]) AND BMJ [All Fields]). There are <90 hits; but searching on **ANGINA AND BMJ [JOURNAL NAME!]** (exactly like that) automatically maps to (('angina pectoris'[MeSH Terms] OR angina[Text Word]) AND (('["BMJ"] [Journal Name] OR "Br Med J" [Journal Name] OR 'Br Med J (Clin Res Ed)' [Journal Name] OR 'BMJ' [Journal Name])) This yields >300 matches—ie MeSH can expand as well as limit your search.

Other useful things to put in square brackets relate to authors [AUTHOR], publication dates [PDATE] and publication types [PTYPE]—eg entering **ANGINA AND RANDOMIZED CONTROLLED TRIAL [PTYPE] AND BLACK DM [AUTHOR, 1996-2005[PDATE]** gives a search which doesn't include articles using common words such as black, and goes from 1996 to now. Using Random* may be more inclusive than using **RANDOMIZED CONTROLLED TRIAL [PTYPE]**, but will include sentences such as 'there are no randomized trials'.

Finding MeSH headings You can make search terms more certain by selecting them via the **MeSH browser** button—look carefully: it's in the left-hand blue margin, half way down. Typing into the search box yields **MeSH terms** which need no mapping. If no exact term is found, choose a likely one from the list offered, and press the 'browse term' button. Click the 'Add' button to add this to your search. Try this with **SMALL VESSEL ANGINA**. This is not a MeSH term, but the MeSH browser offers Angina, microvascular, among a host of other less helpful possibilities. Confirm this for yourself.

Other ways of limiting searches Click the 'Limits' button to explore this, or add a word such as **ABSTRACT** (one word, no space, added to the search phrase) to retrieve only those articles which have abstracts (but this will exclude editorials and correspondence items for example). To exclude **Boonin**, try **MYOINFARCT AND MYOCARDIAL INFARCTION NOT PRINZMETAL ANGINA [NAME]**. This excludes studies of Prinzmetal's angina.

EBM Try **ANGINA AND COCHRANE ANALYSIS [COCHRANE AUTHOR]** or **ANGINA AND META ANALYSIS**, or try the **clinical queries** button. Choose 'sensitive' to avoid missing possible hits, or 'specific' if you are getting too many hits.

Exercises Now you've got the idea, recall your last 6 patients, and find any meta-analyses relevant to them. **Any problems, solutions, suggestions?** • Misspellings? • Not using capitals (AND, OR, NOT)? • Using the wrong search box?

Audit

Audit comes from the Latin *audire*—to hear; and the term was once used for verbally presented verified financial accounts. Audit in clinical practice involves quality control by systematic review of (an aspect of) practice, implementing change and verifying that the desired effect was produced. Its purpose is as a tool to achieve best quality clinical care. Audit means asking questions like: 'Have we any agreed aims in medical practice?' and 'Are we falling short of these aims?' and 'What can we do to improve performance?' Audit is a part of the Summative Assessment required for all UK GP registrars to pass before they can become principals. As from 2002 an 8 point audit is required. [See examples]

- 1 **Title** When selecting a topic think: is it relevant; common or important; measurable; amenable to change? For a registrar audit, is it simple? For other audits, is it worth the investment (of time and money)? Say why you chose the particular topic. (Diabetes: *Diabetes: poor diabetic control*.)
- 2 **What criteria were chosen** State why the criterion/criteria were chosen. (Diabetes: *Diabetes: poor diabetic control*.)
- 3 **Setting standards** A standard is a statement of a criterion of good quality care. A target should be set (the degree to which the criterion will be met). Choose a realistic and obtainable target. Aim for standards to be evidence based. (Diabetes: *Diabetes: poor diabetic control. Target to have >50% patients with HbA1c <7.5%*)
- 4 **Planning and preparation** What have you done? For example, what literature has been consulted (essential). (Diabetes: *Diabetes: poor diabetic control. Target to have >50% patients with HbA1c <7.5%*.)
- 5 **First Data Collection** Gather the evidence. Observe current practice. Compare this to the standard. (*40% HbA1c <7.5%*)
- 6 **Implement changes** (Diabetes: *Diabetes: poor diabetic control*.)
- 7 **Second Data Collection** Compare with the standard and the first data collection. (Diabetes: *Diabetes: poor diabetic control. 45% HbA1c <7.5%*.)
- 8 **Conclusion/Reflection** (Diabetes: *Diabetes: poor diabetic control*.) Use these to formulate your next title to complete the **audit cycle**. (Diabetes: *Diabetes: poor diabetic control*.)

Other people's audits can seem boring. It is only when a practice engages in audit itself that interest is aroused, and it can be satisfying to watch one's practice develop through a series of audits. With computers, audits can be done on many aspects of care, to answer questions such as—Is our care of diabetics adequate? Are all our fertile female patients rubella-immune?

The practice manager can have a central rôle in running an audit exercise—eg by relieving doctors of the burden of data collection, and is able to communicate the results of the audit in a practice's annual report.

Positive aspects of audit (No intervention is without side-effects.)

- It takes time away from eye-to-eye contact with patients.
- In becoming the province of professional enthusiasts, it can alienate some practice members, who can then ignore the results of the audit.
- There is no guarantee that audit will improve outcomes.
- It may limit our horizons—from the consideration of the vast imponderables of our patients' lives in a world of death, decay, and rebirth—to a preoccupation with attaining tiny, specific, and very limited goals.
- Some doctors fear that in espousing audit they risk transforming themselves from approachable but rather bumbling carers and curers who perhaps don't know exactly where they are going, into minor administrative prophets, with too much of a gleam in their eyes and zeal in their hearts.

Appraisal, assessment, and revalidation

► *Appraisal is not the same as assessment.* For NHS UK GPs (for whom yearly appraisal is a contractual requirement) peer appraisal is least threatening when seen as a chat with a colleague about one's professional development.

What form does UK appraisal take? After completing a few administrative forms (Forms 1-3) there is a form (Form 4) requiring a period of thought and data gathering (in protected time). This entails a commentary on your work, an account of how your work has improved since your last appraisal, your view of your continuing developmental needs, and cataloguing of factors which constrain you in achieving your stated aims. Questions include:

- What are the main strengths and weaknesses of your clinical practice?
- How has the clinical care you provide improved since your last appraisal?
- What do you think are the clinical care development needs for the future?
- What factors constrain you in achieving your aims for your clinical work?
- What steps have you taken to improve your knowledge and skills?
- What have you found particularly successful or otherwise about these steps?
- What professional or personal factors constrain you in maintaining and developing your skills and knowledge?
- How do you see your job and career developing over the next few years?
- What are your main strengths and weaknesses in your relationships with patients? How have these improved? What would you like to do better? What factors in the workplace (or more widely) constrain this?
- What are your main strengths and weaknesses in your relationships with colleagues? How have these improved? What would you like to do better? What factors in the workplace or more widely constrain this?
- Do you have any health-related issues which might put patients at risk?

Other areas: teaching; financial probity; research; management activities.

During the appraisal Form 4 is completed and signed by the assessor and the assessee. In anonymized format, Form 4 feeds through to Clinical Governance Leads, who identify trends, and makes a report to the Chief Executive of his or her Primary Care Organization. Form 5 is a non-obligatory form containing background ideas supporting Form 4 which may be used to inform subsequent appraisals. Finally there is a mechanism for supportive follow-up (eg a further visit or phone call).

In the words of the chief medical officer, appraisal is 'a positive and supportive developmental process, a constructive dialogue structured in such a way that those being appraised have the opportunity to reflect on their work and consider any developmental needs. It gives individuals the chance to assess their career path and consider how they might gain more job satisfaction from their current rôle. And by giving feedback on performance it provides the opportunity both to identify any factors that adversely affect performance, and to consider how to minimize or eliminate their effects. It is an important building block in a clinical governance culture that ensures high standards and the best possible patient care.' www.doh.gov.uk/cmo/cmo0203.htm

There is a big question-mark over 'ensures', above. The effect of appraisal on patient care is unknown—but, appraisal, it is hoped (and it only is a hope) can offer opportunities, for interdependent support, self-education, self-motivation, and career development in the wider medical world. It may also be a catalyst for change and even a tonic against complacency. [medline:10822914](http://www.doh.gov.uk/cmo/cmo0203.htm)

Appraisal only makes sense assuming that GPs want to be good, professional, life-long learners. If this is not the case, the less soft realms of revalidation, performance management/assessment will bite. <http://www.appraisals.nhs.uk>

Revalidation (eg every 4–5yrs) gives authority to continue practicing, and to some extent guarantees public safety. Appraisals feed into this process.

Under- and over-performing doctors

It would be nice for the public and the 'leaders' of our profession if there were a small number of underperforming doctors who could be retrained or struck off. Things are rarely so simple, and we may have to accept that, for many reasons, including chance, training, and resilience, the performance of *all doctors* will, at times, be, or appear to be suboptimal. If all doctors were the same, and there was a valid yardstick for measuring quality (a big 'if'), then there would be, by chance, a large underperforming group, with a corresponding apparently 'over-performing' group. Anyone assessing performance data needs to take into account these questions:

What counts as data? Usually only what can be quantified. Quality is different.

Is the data stratified for risk? Doctors' case-mixes can vary markedly.

Has our personal data's accuracy been validated? Data entry is often unreliable.

Has the accuracy of the data we are being compared with been validated?

Could the differences between our data and others have arisen by chance?

This is the most revealing question. Imagine a thought-experiment in which 4 equal doctors use different strategies for predicting whether a tossed coin will land heads or tails. One always chooses heads, one always chooses tails, and the other two alternate their choices out of synchrony with each other. When I did this experiment for a pre-decided 14 throws each (56 throws in total), the best doctor only had 2 errors, whereas the worse had 7 errors—over 3 times the rate for post-operative deaths (or whatever). The public would demand that this doctor be retrained or struck off, and the General Medical Council might feel obliged to comply, simply to keep public confidence (it is under great pressure to 'do something'). So must we all be prepared to be sacrificial lambs? The answer is Yes, but there are certain steps that can be taken to mitigate our own and our patients' risk-exposure.

- When we encounter doctors who are clearly underperforming (eg due to addictions) we must speak out. This will encourage belief in the system.
- For statistical reasons any series with <16 failures might be best ignored. Such series simply do not have enough power to detect real effects.
- We must strive to be both kind and honest with our patients. The best response to 'I'd like a home-delivery' might be to say 'I haven't done one for 5 years—and that one went wrong: are you sure you want my services?'—rather than 'the uk perinatal death-rate is the same for home and hospital'.
- It might be the case that, contrary to the GMC, we should *not* always be on the look-out for colleagues who might be underperforming so that we can report them to the proper authorities: rather we should be encouraging an atmosphere of mutual support and trust—the sort of environment in which doctors feel safe to say 'all my cases of X seem to be going wrong—can anyone think why?' To stop this trust turning into cronyism we must be prepared to engage in, or be subjected to, audit (p 448). The alternative is for clinicians to develop into secret police, informers, and counter-informers. No one would benefit from this. We note that malicious informing is not an isolated occurrence in the uk; 80% of those suspended for presumed underperformance are exonerated, but few return to their previous job owing to the stresses enquiries always engender.

Typical areas in which doctors are seen to be underperforming

Local Medical Committees (LMCs), complaints bodies, and NHS commissioners have all been systematically questioned about doctors whose performance they are reviewing. In the case of LMCs, for example, clinical skills were the chief worry, followed, in order, by communication problems, management problems, prescribing problems, and record-keeping problems. NHS Trusts have more concern over referral patterns.

Patient satisfaction

► *The patient is the nearest thing we have to an infallible judge of what constitutes good medicine.* In summary, what patients mostly want is a *personal service* from a *sympathetic doctor* who is *nearby* and *easy to get to see*. Satisfaction is one of the few measures of *outcome* (not *process*) which is fairly easy to measure, eg by questionnaire agreement with these 13 statements (*Baker's dozen*¹).

Satisfaction

- 1 I am totally satisfied with my visit to this doctor.
- 2 Some things about my visit to the doctor could have been better.
- 3 I am not completely satisfied with my visit to the doctor.

Professional care

- 4 This doctor examined me very thoroughly.
- 5 This doctor told me everything about my treatment.
- 6 I thought this doctor took notice of me as a person.
- 7 I will follow the doctor's advice because I think he/she is right.
- 8 I understand my illness much better after seeing this doctor.

Relationships

- 9 This doctor knows all about me.
- 10 I felt this doctor really knew what I was thinking.
- 11 I felt able to tell this doctor about very personal things.

Perceived time

- 12 The time I was allowed with the doctor was not long enough to deal with everything I wanted.
- 13 I wish I could have spent a bit longer with the doctor.

Why do patients change their doctor? The most common reasons are that either the patient has moved, or the doctor has retired or is perceived to be too far away. There are additional reasons:²

Patient needs:		Organizational problems:		Problems with doctor:	
One doctor for all the family	5%	Long waits	13%	Lost confidence in	21%
Want woman doctor	4%	No continuity of care	6%	Uninterested/rude	20%
Want alternative medicine	2%	Rude receptionist	6%	Prescriptions criticized	5%
Obstetric needs	1%	Appointments wanted	1%	Doctor too hurried	4%
		Open surgeries wanted	1%	Visits problematic	4%
		Other staff rude	1%	Communication poor	4%

A USA study found that a *participatory decision-making* style leads to patient satisfaction. Participation was found to depend, in part, on the *degree of autonomy* perceived to be enjoyed by the GP, and on the *volume of work*.³

Another approach to gaining satisfaction is to agree and publish standards of care that patients can expect, along with performance figures for how nearly these standards are achieved in practice. This is the philosophy behind the UK government-led *Patient's Charter* and the British Standards kitemark BS5750—the aims of which are to:

- Set standards, eg by agreement with patient participation groups (p 440).
- Monitor progress towards these standards, and publish progress locally.
- Provide information about how services are organized. Maximize choice.
- Let users know who is in charge of what, and what their rôles are.
- Explain to users what is done when things go wrong, and how services are improved, and what the complaints procedure is.
- Show that taxpayers' money is being used efficiently.
- Demonstrate customer satisfaction.

This culture has proved alien to most GPs, perhaps owing to a very necessary pre-occupation with illness and its curing, rather than service, and its glorification.

1 R Baker 1990 *BJGP* 487 & 1996 601 2 B Billingham 1993 *Ibid* 43 336 3 S Kaplan 1996 *An Int Med* 124 497

Stopping smoking tobacco

see www.nice.org.uk (2002)

Epidemiologists estimate that ~50% of smokers will die of smoking if they do not give up—and it is not the case that they only lose a few years: a quarter of a century is more likely for those dying between the ages of 35 and 69.¹ Stopping smoking diminishes the excess risk from tobacco, so that after 10–15yrs the risk of lung cancer effectively reverts to that of lifelong nonsmokers. A similar, but quicker diminution of excess risk (\downarrow by ~50% in the first year) is found for deaths from coronary artery disease and, to a lesser extent, risk of stroke. *~60% of smokers want to give up.*

Annual UK health costs of smoking

- GP consultations: >£89 million
- GP prescriptions: >£52 million
- Hospital episodes: >£470 million

Advantages of stopping smoking Saving of life (110,000/yr in UK).

- Larger babies (smokers' babies weigh on average 250g less than expected, and their physical and mental development may be impaired).
- Less bronchitis (accounts for millions of lost working days).
- Less risk from the Pill: cardiovascular risk $\uparrow \times 20$ if uses >30 cigarettes/day.
- Less risk from passive smoking (cot deaths, bronchitis, lung cancers).
- Return of the sense of taste and smell—and relative wealth.

Helping people quit (AAAA = ask, advise, assist, arrange follow-up.)^{1,3,4}

- Ask about smoking in all consultations, not just those on smoking-related diseases (be subtle; patients won't listen if you do not share agendas).
- Advise according to need. Ensure that advice is congruent with beliefs; motivate patients by getting *them* to list the advantages of quitting.
- Assist in practical ways, eg negotiate a commitment to a 'quit date' when there will be few stresses; agree on throwing away all smoking junk (cigarettes, ash trays, lighters, matches) in advance. Inform friends of new change.

Nicotine gum, chewed intermittently to limit nicotine release: \geq ten 2mg sticks may be needed/day. Transdermal nicotine patches may be easier. A dose increase at 1 week can help. Written advice offers no added benefit to advice from nurses/GPs. Review at 2 weeks; people sense (and act on) your commitment. Only re-prescribe if abstinent.

Bupropion ups the quit rate to perhaps 30% at 1yr vs 16% with patches: consider if the above fails. Dose: 150mg/24h po (while still smoking; quit within 2 weeks); the dose may be doubled from day 7; stop after 7 weeks. Give enough to last 3 weeks, then review; only re-prescribe if abstinent—ie the *transdermal nicotine patch* regimen.⁵

Contraindications: Seizures (risk <1:1000), insomnia, headache, arrhythmias.

Contraindications: Epilepsy; cirrhosis; pregnancy; lactation; bipolar depression; eating disorders; CNS tumours; alcohol/benzodiazepine withdrawal; ≤ 18 y old.

Contraindications: Antimalarials, antipsychotics, antidepressants, antihistamines, antiepileptics, quinolones, theophylline, tramadol, steroids.

For those who do not want to give up Give them a health education leaflet, record this fact in the record, and try again later.

~25% of school leavers smoke regularly. The Health Education Council has a smoking education project for schools. It has been commented that smoking rates may not be rising too fast in children owing to cost, and to mobile phones, which are cheaper, and just as good a fashion accessory.

Policy implications should have a policy on promoting nonsmoking, and offering practical advice: primary health care teams; hospitals; midwives; pharmacies—and also, perhaps, schools and employing authorities. Health commissioners also needed to promote knowledge and training in this area.¹

Reducing alcohol intake

With the toll that excess alcohol takes in terms of personal misery and the national purse (>£1600 million/yr in the UK), the need to reduce alcohol intake should rank as one of the leading aims of preventive care. The reason why alcohol is not at the top of the agenda is not just that doctors are so fond of it (the profession has three times the national rate of cirrhosis), but because there is a powerful and pervasive lobby which ensures that alcohol is cheaper (in relative terms) and more readily available than ever before—so that its use on an individually moderate scale arouses no comment. It is assumed to be safe, provided one is not actually an alcoholic. However, it is more helpful to view alcohol risks and benefits as a spectrum (see the French Paradox, *OHCM* p 194 for the benefits of alcohol). Problems are listed on p 363. A strategy to reduce the bad effects of alcohol in your patients:

- Whenever a patient presents with symptoms or signs related to alcohol, be sure to ask in detail about consumption.
- Question any patient with 'alerting factors'—accidents, driving offences, child neglect, assault, attempted suicide, depression, obesity. Question others as they register, consult, or attend for any health check.

Time interventions Time interventions for when motivation is maximal, eg as (or before) pregnancy starts. Small reductions *do* matter.¹

- Take more nonalcoholic drinks.
- Limit your drinking to social occasions—and learn to sip, not gulp.
- Reduce the frequency of sips, eg by shadowing a slow drinker in the group. Don't pick up your glass until he does (and don't hold your glass for long; put it down to avoid unconscious sipping).
- Don't buy yourself a drink when it is your turn to buy a drinks' round.
- Go out to the pub later. Take 'days of rest' when no alcohol is drunk.
- Learn graceful ways of refusing: 'No more for me please, I expect I'll have to drive Jack home' or 'I'm seeing what it's like to cut down.'

Planning reduced drinking • Agree goals with the patient.

- Suggest he keeps an alcohol diary in which he records all drinking.
- Teach him to estimate his alcohol intake (u/week, see below).
- Consider an 'Alcohol Card' in the notes to show: units/week; pattern of drinking; reasons for misuse; each alcohol-related problem (and whether a solution has been agreed and action implemented); job record; family events; biochemical markers (GGT, mcv); weight.
- Give feedback about how he is doing—eg if GGT (γ -glutamyl transpeptidase) falls are discussed at feedback, there is much lower mortality, morbidity and hospitalization compared with randomized control subjects.
- Enlist family support; agree a system of 'rewards' for sobriety.
- Group therapy, self-help groups, disulfiram, local councils on alcohol, community alcohol teams and treatment units may also help (p 363).

Setting limits to low risk drinking eg ≤ 20 u/week if σ ; ≤ 15 u/week if φ —there are no absolutes: risk is a continuum. NB: higher limits are proposed, on scant evidence¹ (eg 4u/day; 3u for women). One unit is 9g ethanol, ie 1 measure of spirits, 1 glass of wine, or half a pint of ordinary-strength beer.^{2,3} *Primary care is a good setting for prevention*: intervention leads to less alcohol consumption by ~15%, reducing the proportion of heavy drinkers by 20%—at one-twentieth the cost of specialist services.⁴ There is no evidence that GP intervention has to include more time-consuming advice such as compressed cognitive/behavioural strategies.^{5,6} Simple advice works fine as judged by falling GGT levels—at least for men. After interventions, women may report drinking less, but this is not reflected in a falling GGT.⁵

Domestic violence^{1,2}

This is common and includes physical, sexual, emotional, and psychological abuse. It is rarely an isolated event, and often escalates in severity and frequency. ► Do not hesitate to ask directly about this in consultations

Epidemiology Much domestic violence is unrecognized and is probably associated with alcohol or substance abuse. It is estimated that:

- One in three women experience domestic violence at some time in their lives (perhaps 1 in 10 in the last year).
- About 30% of domestic violence starts in pregnancy.
- It can often escalate during pregnancy or after childbirth.
- Over a million incidents of domestic violence are recorded by the police each year (1 in 4 of all assaults).
- Of women murdered, 40% are killed by a current or ex-partner.

Those who are abused are:

- 5 times more likely to abuse alcohol.
- 9 times more likely to abuse drugs.
- 3 times more likely to be diagnosed psychotic or depressed.
- 5 times more likely to commit suicide.
- 15 times more likely to suffer a miscarriage than non-abused women.

Those who work in the health service should be aware that abuse is common. Those who are abused may attend frequently with trivial or non-existent complaints; or conversely not attend for treatment because of lack of money. They may minimize signs of violence on the body; be evasive or reluctant to speak in front of partners, and partners may tend to be constantly present so that it may be difficult to talk to the woman alone. This can be particularly difficult if the partner is required for translation purposes: here another translator should be sought who is not from the family.

Social Services (and others) should be prepared to help women disclose domestic violence (for example, whenever there is injury or unexplained phenomena—and it is now recommended that abuse be enquired about at routine antenatal bookings so that issues can be addressed *before* injury and fatalities occur).¹ Social Services may be in a position to provide support and practical advice about options available and should be prepared to offer support over long periods. Involve social services if children's welfare is involved (see p 284).

Refuge The Women's Aid Federation (0345 023468^{UK}) can provide legal advice, emotional support, and refuge. The police may also help.

Court orders¹ These may be obtainable quickly, and may be the only way to prevent someone going near a previous victim. They may also require someone to leave a place of residence, or let the victim return home. They are not a long-term solution, but may be lifesaving in emergencies. In the UK, Law Centres exist to provide access to legal protection: to find your nearest one, phone 0207 387 8570. Alternatively, the patient's solicitor may be available. The police may also need to be called.

See also *Child abuse*, p 284–5.

1 DoH 1998 *Confidential Enquiries into Maternal Deaths*, Chapter 13

2 A Sandell 1998 *Oxford Handbook of Patient's Welfare*, OUP www.oup.co.uk/OHPW

Alternative and holistic medicine

1 We need to know about alternative medicine to understand our patients undeclared distress, which use of nonstandard treatments is so often a sign of.¹ We also need to be able to advise on the safety of various therapies.

'Alternative' describes therapies which are not included in conventional medical/paramedical training—although an increasing number are being subjected to rigorous assessment in double-blind, randomized trials.²

Some are the orthodoxies of a different time (eg *herbalism*) or place (the *Ayurvedic medicine* of India), some are mainly diagnostic techniques (*iridology*), some therapeutic (*aromatherapy*). Some doctors are suspicious of unorthodox medicine, and feel that its practitioners should not be 'let loose' on patients. But in many places the legal position is that, however unorthodox a practitioner's treatment may be, he or she cannot be convicted of unethical practice in the absence of clear harm to patients.²

Many people (~5 million/yr in the UK) consult alternative practitioners, usually as a supplement to orthodox treatment and for a limited range of problems—predominantly musculoskeletal. Many will feel unable to tell their doctor about trips to alternative therapists, unless asked.

Modern medicine is criticized (both from within and from without) for sacrificing humanity to technology, and with little benefit for many patients. In contrast to the orthodox doctor, alternative practitioners may be seen as taking time to listen, laying on hands rather than instruments, and giving medicines free (not always!) from side-effects.

Many alternative therapies seem implausible, and controlled trials are hard, if treatment is individualized and therapists' personality is a vital factor. But the same comments could be made of much orthodox care.

Acupuncture Traditionally used to treat a wide range of conditions, acupuncture is increasingly employed in orthodox practice for pain relief, control of nausea and treatment of addiction. For these, endorphin release provides a scientific rationale.

Homeopathy This is based on the principles that like cures like, and that remedies are made more efficacious ('potentiated') by infinite dilution. UK NHS GPs may prescribe homeopathic remedies on an FP10. Randomized trials suggests real (small) benefits, eg in asthma, but nobody knows if the reason why they are effective is 'like cures like'.^{3,4}

Yoga and other therapies *Ashtanga yoga* and *therapeutic yoga*. These are widely used and of proven benefit in musculoskeletal problems, but some use them to treat more general conditions such as asthma.⁵

Clinical ecology Starting from the fact that atmospheric pollutants, toxins, and xenobiotic chemicals (from other organisms) are known to be harmful, a system is built up around techniques (using intradermal injections) for provoking and neutralizing symptoms related to foods.^{2,5}

Holistic medicine Holism entails a broad view: of the patient as a whole person, of the rôle of the therapist, of the therapies used. The patient's autonomy is encouraged through involvement in decisions, and nurturing of self-reliance. • *Specialism doesn't exclude holism*: nephrologists can be as holistic as naturopaths. As shown on p 422, most models of the GP consultation are based on a patient-centred approach which is essentially holistic. Compare the sequence 'bronchitis → antibiotic' with 'bronchitis → smoker → stressed → redundancy-counselling → ?antibiotic'.

Living dangerously

Some years ago a patient had a seminoma treated, apparently successfully, in a well-known London hospital, to which he had been referred from his distant Sussex village. In the year that follow-up stopped, the patient had a major myocardial infarction—again, followed by an apparently reasonable recovery. But the patient became morbid, self-centred, and depressed, perhaps because of the dawning appreciation of his mortality, his residual breathlessness, and his inability to carry out his hobby of carpentry. His GP tried hard to cheer him up, and rehabilitate him by encouraging exercise, sex, a positive self-image, and alternative hobbies. Rehabilitation was almost working when he began to develop headaches and kept asking forlornly whether these were a sign that his cancer had spread to his brain. There were no signs of recurrent tumour or raised intracranial pressure. His GP appreciated that there was a chance that the tumour was resurfacing, but judged that starting a pointless chain of investigations would be disastrous to the patient's mental health. So instead of arranging CT scans the GP interpreted the patient's forlorn question for him, saying that he was only asking questions like this because he was in a negative frame of mind, and the patient and his GP developed strategies to avoid negative cognitions, in co-operation with the local consultant who had helped to look after his myocardial infarction. The headaches improved, and the pressure to investigate was resisted.

Had a CT scan been done, it would by no means have achieved reassurance if it was negative—in the patient's frame of mind he would be all too willing to ask if the CT scan is 100% reliable, and then to request some other test in addition, and so on, until illness had become a major preoccupation. So in this case, it was rational for the doctor to live dangerously, take risks, and be prepared to take the blame if things had gone wrong.

How do we cope with and thrive on uncertainty? The first step is to get away from the idea that if you do not do all you can to reduce uncertainty, you are somehow being lazy. (The reverse may be true.) The next step is to share the uncertainty with a colleague—to see if he or she agrees with your judgment. From the medicolegal viewpoint it is wise to document your thought processes. Another caution is to follow in the steps of those adventurous but wise mountaineers who never plan a route without also planning an escape route: in the medical sense, this means the triad of follow-up, the taking of the family into one's confidence, and honest reflection on the chances of error and the chances of detecting it. This means that, as far as possible, you will get early warning of error, and then be able to adjust your therapeutic approach in line with the way the illness unfolds.

Minor illness

Many people with apparently minor illnesses visit their GPs, although GPs do not have a sufficient monopoly of this to justify being called 'triviologists'. Most minor illnesses are dealt with from a disease point of view, in the relevant chapter in which they occur. Here, the concern is with the study of minor illness itself, and in this context much minor illness does not come to the general practitioner: only people come to general practitioners, and it may not be known for some time whether the symptoms are serious or minor (minor to whom?). A GP may not want to spend all his time on minor conditions, but this may become almost unavoidable if he issues a prescription for such complaints. This reinforces attendance at the surgery, as a proportion of patients will come to assume that a prescription is necessary. In current practice, GPs rate about 14% of their consultations as being for minor illness (mild gastroenteritis, upper respiratory problems, presumed viral infections, flu, and childhood exanthemata).¹ More than 80% are likely to receive a prescription, and >10% are asked to return for a further consultation. Why does this great investment of time and money occur? Desire to please, genuine concern, prescribing to end a consultation, and therapeutic uncertainty may all play a part. Positive correlations with low prescribing rates include a young doctor, practising in affluent areas and long consultation times. Patients in social classes I and II are more likely to get a home visit for minor ailments than those in other social classes. Membership of the Royal College of General Practitioners does not influence prescribing rates.¹ Not everyone wants to reduce prescribing, but advice is available for those who do:²

- Using a self-care manual explaining about minor illness.³
- Using self-medication (eg paracetamol for fever).
- Using the larder (eg lemon and honey for sore throats).
- Using time (eg pink ear drums⁴—follow-up).
- Using granny (a more experienced member of the family).
- Pre-empting the patient's request for antibiotics (eg for a sore throat), eg: 'I'll need to examine your throat to see if you need an antibiotic, but first let me ask you some questions. . . From what you say, it sounds as if you are going to get over this on your own, but let me have a look to see.' [GP inspects to exclude a quinsy.] 'Yes, I think you'll get over this on your own. Is that all right?'

Empowering patients Any illness, minor or otherwise, is an opportunity to empower patients. Use the time to enable patients to improve their ability to:

- Cope with life.
- Understand their illness.
- Cope with specific illnesses.
- Feel able to keep themselves healthy.
- Feel confident on handling health issues.
- Be confident about the ability to help themselves.

We know that time spent this way improves patient satisfaction and clinical outcome.^{5,6}

Are you ready to be on call tonight?

In your black bag (Keep in date!) Contents depend on local needs, eg:

Minor agents	Older agents	Emergency agents
Netilmicin; ceftriaxone	Pain killers; antibiotics	Mental Health Act forms
Cyclimorph®; naloxone	Prednisolone (soluble)	Headed notepaper, etc.
Prochlorperazine	Lofepamine or SSRI	Phone N°: chemists, police
Frusemide	Ranitidine or similar	ambulance, hospitals, etc.
Atropine; adrenaline	Aspirin 75 & 300mg	Certificates; prescriptions
Chlorpheniramine	Paracetamol 'syrup'	inc. nursing for iv pumps
Benzylpenicillin	Rehydration sachets	Claim forms: temporary
Diazepam	Inhalers; GTN spray	resident; immediately
Glucagon	Enemas & suppositories	necessary treatment
50% glucose	Fucithalmic ointment	Book to record batch num-
Haloperidol	Hypostop® glucose gel	bers for ampoules & nar-
Thrombolysis	Diazepam (pills + PR)	cotic use <small>medline:11064942</small>

The suppositories referred include diclofenac 100mg (eg for renal colic) and paracetamol 60mg or 125mg for vomiting feverish children.

Equipment Airway; stethoscope; auroscope; patella hammer; scalpel blade; BP wrist monitor or anaeroid device; FeverScan® (or similar, for T°; no mercury); dipstix/capillary glucose; needles; syringes; gloves/kv jelly®; antiseptic fluid; sutures; specimen bottles & forms; sharps tin.

► This is just what is in the author's bag: it is not prescriptive.

In your pocket Bleep/phone (charged!). Try a **palmpc device** with this book (we declare an interest!), *OHCM & BNF/pharmacopoeia*; see www.franklin.com
NB: the trouble with relying on books is that there is no room for more than one in a bag, and the danger is that if you keep them in your car, they are not to hand for easy reference. No-one wants to get to the top of a block of flats only to have to descend to see if x interacts with y or to find out the incubation time for scarlet fever. **Permanent, or internet access** (eg with an xda® device*) means that as well as locally-loaded books, everything else is available too in the way of decision support. For example, www.isabel.org can be very helpful with paediatric problems. In due course, these devices will also be able to access patients records remotely.

In your car Maps, nebulizer, spare batteries, speculum, defib...?ECG, dressings, giving sets for ivis; peak flow meter; O₂.

In your mind We have a duty to be fully conscious and reasonably healthy. We have all been in the position of visiting patients who are less sick than we are—and we tend to carry on until we drop (see p 605). This is bad for us and bad for patients. If you are sick, and no locum is to hand, phone your local Primary Care Organization or out of hours co-operative.

*This is an O₂-services integrated phone and handheld computer, using GPRS (general packet radio service)—so e-mail is instant. www.mmo2.com/docs/services/xda_details.html
 • FujitsuSiemens Loox PC www.expansys.com/product.asp?code=FSLOOX
 (batteries are rechargeable: operating time: standby: ~300h; operating ≤12h) • Toshiba e740 pocket PC: www.expansys.com/product.asp?code=TOSH_E740 • www.palm.com

Prescribing and compliance ('concordance')

On any day ~60% of people take drugs, only half of which are prescribed. The others are sold over the counter (otc). The commonest otc's are analgesics, cough medicines, and vitamins; for prescribed drugs the common groups are CNS and cardiovascular drugs, and antibiotics. On average, 6–7 NHS prescriptions are issued/person/year (21 in Italy and 11 in France).

GPs account for 75% of NHS annual prescribing costs (>£2000 million, or ~10% of the total cost of the NHS), although many of these 'GP drugs' will have been initiated in hospital. The cost of these prescriptions has risen by a factor of ~5 since 1949 (after allowance for inflation) and is ~£100,000–£300,000/GP/year. Positive correlations with low prescribing include a young doctor, practising in an affluent area, and longer consultation times. The reason may be that if extra time is spent with the patient, more explanation about minor ailments (p 458) may be given, so that a patient's expectation for a prescription is replaced by enlightened self-awareness.

Formulary aim is to reduce drug costs and to make prescribing more effective, by producing an agreed list of favoured drugs. This voluntary restriction can work in tandem with compulsory NHS restricted lists, and lead to substantial savings (eg 18%). The UK DoH recommends development at individual practice level but this time-consuming task may be better achieved by adapting an existing local formulary. Unless you wish to reinvent the wheel, this would seem excellent advice. But beware: denying GPs drug choice may increase referral to secondary care, leading to *increased costs*. medline:11735668

Dispensing doctor Where there is no chemist's shop (eg rural areas) GPs are paid to dispense to their patients. Their annual prescribing rate can be as low as 70% of their non-dispensing fellow GPs.

Compliance (Does the patient take the medicine?) *There is no point in being a brilliant diagnostician if nobody can be persuaded to take your treatments.* Even in life-threatening conditions, compliance is a major problem occurring in up to 56% of patients (eg adolescents with acute lymphatic leukaemia). The following are associated with increased compliance:

- Being able to identify with a personal doctor.
- Patient's overall satisfaction with the doctor.
- Simple therapeutic regimens.
- Written information (use short words—Flesch formula >70, *OHCM* p 3).
- Longer consultation times or prescribing on home visits.
- Prescribing in association with giving health education.
- Continuity of care, coupled with belief in efficacy of the treatment.
- Short waiting time for appointments.
- The encouragement of self-monitoring by the patient.

Monitoring Monitoring plasma drug levels is the most reliable way of doing this, but it is cheaper to ask patients to return with their tablets, so that you can count them—or, better still, establish a basis of trust so that the patient can check for him- or herself.

Concordance or concordance of Compliance suggests that you know best and patients who lapse are foolish. But it is known that patients who adapt advice to their needs get fewer side-effects, eg GI bleeding: your prescription may read 'ibuprofen 400mg/8h'—but the patient may, sensibly, only take the medicine when his joints are bad. Don't think of this as the patient failing to do something. It is you who have failed to reach a shared understanding of the pros and cons of medicine-taking with your patient. Concordance denotes even more than this: think of it as a liberating concept, promoting egalitarianism within medicine. *There is no higher ideal.* medline:10181503

Healing

Since neolithic times, healing has had a central place in our culture, and has long been recognized as 'mor bettir and mor precious þan any medicyne' (*OED* v 152.1). Recently medicines have improved greatly, so that the rôle of doctors as the purveyor of medicines has eclipsed their more ancient rôles. We all recognize the limits of our rôle as prescribers, and we would all like to heal more and engage in repetitive tasks less often. But what, we might ask, is healing? How is it different from curing? Healing is, at one level, something mysterious that happens to wounds, involving inflammation→granulocyte, macrophage, and platelet activation→release of platelet-derived growth factor & transforming growth factors α and β →neovascular growth→fibroblast-mediated contraction→proteoglycans and collagen synthesis, lysis, and remodelling (see *Oxford Textbook of Surgery*).

On another level, healing involves transforming through communication—a kind of hands-on hypnosis. We can cure with scalpels and needles, but these are not instruments of communication. Here is an example of healing (an all too rare event in our own practice). On a rainy February evening, after a long surgery, I visited a stooped old man at the fag-end of life, with something the matter with his lung. 'I suppose it's rotting, like the rest of me—it's gradually dying.' I reply: 'Do you think you're dying?' 'Aren't we all?' 'Green and dying' I reply for some reason, half remembering a poem by Dylan Thomas (*Fern Hill*). The patient looks mystified: he thinks he misheard, and asks me to repeat. 'Green and dying' I say, feeling rather stupid. There is a pause, and then he rises to his full height, puffs out his chest, and completes, in a magnificent baritone, the lines: '... Time held me green and dying, though I sang in my chains like the sea.'

www.breakoutofthebox.com/fernhill.htm

By chance I had revealed a new meaning to a favourite poem of his which perhaps he thought was about childhood, not the rigours of his old age. Both our eyes shone more brightly as we passed to the more prosaic aspects of the visit. This illustrates the nature of healing: its unpredictability, its ability to allow us to rise to our full height, to sing, rather than mumble, and how externally nothing may be changed by healing, just our internal landscape, transformed and illuminated. It also shows how healing depends on communication, and how it is bound up with art. Healing may be mysterious, but it is not rare. We have so often kissed the grazed knees of our daughters that we expect the healing balm of kisses to wear out, but, while they are young, it never will, because children know how to receive but not how to doubt, and the kiss is the paradigm of healing: contact between two humans, wordless service of the lips, which the sterile advice of a microbiologist would never condone, an activity for which there is no Read code, and which we know as healing.

Our bread-and-butter work comprises the sifting of symptoms, deciding what is wrong, and prescribing treatment—all tasks which, according to a historic prediction by no less a personage than the editor of the *Lancet* are destined for delegation to microchips.

medline:10836918

This implies that our chief rôle will be as healers and teachers. There is much to be said for the idea that we should throw away the paraphernalia of mechanistic medicine, our formularies, our computers, and our audits—and insist on returning our primordial rôle. Meta-analyses of randomized healing trials (prayer, mental/ spiritual healing, therapeutic touching) bear this out to some extent: 57% of randomized trials show a positive effect. medline:10836918

No doubt there will always be some way to go before healing, the central ideal of medicine, becomes its central activity. After all, the last thing any of us wants, when struck down by appendicitis, is a poet or a healer—but last things will always retain their power to set us thinking.

Social matters

Unemployment and families UK data show an association between child deaths and unemployment, lower social class and overcrowding. Babies whose fathers are employed are heavier at birth (by 150g) than unemployed fathers' babies, after adjusting for other factors. Accidents and infection are more rife among children of the unemployed compared with carefully selected controls, and their mothers may be more prone to depression. As unemployment rises, so does child abuse. Other factors identified with this rise are marital discord, debt, and parents' lack of self-esteem, as affected families reveal: 'When he lost his job he went bonkers. He changed completely. He became depressed and snappy. Frustrated.'

Marital breakdown heads the list of problems of women with neurosis, coming 2nd (to employment difficulties) in men, and is a leading factor in >60% of suicide attempts. In the USA, divorced males have the highest rates of mortality. The greater incidence of cardiac deaths is most marked in young divorced males. Being divorced and a nonsmoker is nearly as dangerous as smoking a pack a day and staying married. Marital harmony (eg cuddling) protects from cardiac death, as shown in one prospective study of 10,000 Israeli hearts. Parental behaviours predicting problematic marriages among offspring included jealousy, being domineering, getting angry easily, being critical, moody, or taciturn.

medline:11642350

Social security benefits For **Child Tax Credit**, see www.dss.gov.uk/lifeevent/benefits
www.dss.gov.uk/lifeevent/benefits/social_security_agreements.htm#azlisted

Child Tax Credit Worth applying for if a pension is the only income.

Child Tax Credit The maximum help is £314 for residential homes and £387 for nursing homes. This type of care is free in Scotland.

Child Tax Credit is available if 3–65yrs old; it is paid if a client needs help looking after himself. It is paid at different rates depending on circumstances. For those <16yrs, it is paid for a child with a physical or mental illness or disability if they need more help or looking after than other children of the same age because of their illness or disability—weekly amounts: *for care*: higher rate=£53.55; middle rate=£35.80; lower rate=£14.20; *for mobility*—higher rate=£37.40; lower rate=£14.20.

Child Tax Credit is paid to help working disabled people on a low income. Savings >£16,000 usually preclude payment. Savings >£3000 affect how much is payable.

Child Tax Credit is paid by local councils, irrespective of other benefits.

Child Tax Credit Tax-free weekly benefit for people aged 65 or over who need help with personal care owing to illness or disability. It is ignored as income for working out Income Support and Jobseeker's Allowance. It is payable to the person needing attending to, and not the person attending. Depending on the disability, the weekly rate is either £37.00 or £55.30. Special fast-track rules apply if terminally ill. The AA stops 4 weeks after going into a home run by the local council or if social services helps with the cost of an independent home.

Child Tax Credit (if aged 16–65yrs) is payable to a person who spends >35h/wk looking after someone who is getting (or waiting to hear about) AA, *Disability Living Allowance* at the middle or highest rates, *Industrial Injuries Disablement Benefit* *Constant Attendance Allowance*, or *War Pensions Constant Attendance Allowance* at the basic rate or above.

To find a patient's nearest **Child Tax Credit**, see www.dwp.gov.uk/localoffice/index.htm

Health and social class

Throughout human history there have been inequalities in the health of classes and populations, caused by social factors. With the introduction of the British National Health Service, with its ideal of equal access to medical care for all groups in society it was assumed that differences in the health of different social (occupational) classes would be eliminated. We now know that this has not happened, and this has been amply documented in various reports such as the *Black Report* (*Inequalities in Health*) and the *Health Divide*. *National Service Frameworks (NSFs)* eg for care of those with ischaemic heart disease, diabetes, mental health problems and for older people who are designed to redress these inequalities.

Registrar General's scale of 5 social or occupational classes

Class I	Professional	eg lawyer, doctor, accountant
Class II	Intermediate	eg teacher, nurse, manager
Class III ^N	Skilled nonmanual	eg typist, shop assistant
Class III ^M	Skilled manual	eg miner, bus-driver, cook
Class IV	Partly skilled (manual)	eg farmworker, bus-conductor, or packer
Class V	Unskilled manual	eg cleaner, labourer

There is a remarkable concurrence of evidence concerning the factor by which mortality rates are higher in social class V compared with those of social class I (with regular gradations between). For stillbirths, perinatal deaths, infant deaths, deaths in men aged 15–64 and women aged 20–59 this factor is respectively 1.8, 2, 2.1, 2, and 1.95. The same sort of factors hold true for specific diseases such as the standardized mortality ratios (SMR) from lung carcinoma (1.98), coronary heart disease (1.3) and cerebrovascular disease (1.9). Only malignant melanoma, anorexia/bulimia nervosa, eczema, and Hodgkin's disease in early adulthood show a reverse ('disease of affluence') trend. Note: the SMR is the ratio of mortality rates in one class compared with the average for the whole population. The whole population has an SMR of 1.00.

There is more emotional stress and chronic ill health in the 'lower' occupational classes. Furthermore, people who own their own homes have less ill health than private tenants, especially if they live in residential retirement areas.

Within occupations the effect of social class is seen in a 'purer' way than when groups of many occupations are compared: in a study of >17,000 Whitehall civil servants there was a greater than 3-fold difference in mortality from all causes of death (except genitourinary diseases) comparing those in high grades with those in low grades. Similarly in the army there is a 5-fold difference in mortality rates from heart disease between the highest and the lowest ranks.

It has been pointed out that being ill makes a person 'descend' the social scale, but it is estimated that this effect is not large enough to account for the observed differences between classes. It is much more likely that the differences are due to factors such as smoking behaviour, education, marital status, poverty, and overcrowding.

Purchasers and providers

2 sets of contrasting principles ▶ He who pays the piper, calls the tune.
▶ Priceless therapeutic assets cannot be bought or sold: these include compassion, continuity of care and commitment. ▶ My job is to spend, spend, spend, until all my patients are healthy. ▶ The job of the Treasury is to squeeze, squeeze, squeeze, until all spending is within government targets. (The clarity of this dichotomy becomes turbid when the doctor is asked to do the squeezing.)

Never just ask how good a structure is without also asking how good it is at transforming itself: that which cannot transform, dies. The UK National Health Service is the largest employer in the Western world and for years the search has been on to find ways to control and transform this dear, mighty thing. The purchaser-provider split is the most powerful lever yet developed for this purpose. Purchasers commission care by drawing up contracts with competing providers, who deliver the care. In England, purchasers are chiefly Primary Care Trusts (which are run by a board comprising a minority of GPs). The better the provider delivers secondary care (do not pause to ask what 'better' means: speculation on this point might ruin the argument) the more likely they are to get the contract next year. The catch is that all the extra effort the provider makes to out-perform a contract this year will probably be taken for granted next year. The same may hold true if purchasing is used for the imposition of guidelines ('evidence-based purchasing'). What has been created is a treadmill which goes faster and faster, while taking less and less account of individual patients' and doctors' legitimate but varying needs. Unless the market is rigged, natural selection ensures that the fittest and fastest providers survive. Patients and taxpayers benefit—until the point where cynicism and exhaustion set in. There is no evidence that once the purchaser-provider path is chosen then cynicism and exhaustion inevitably follow, and there is evidence at local level that benefits accrue, and services become more tuned to consumers' desires. (Consumers are not infallible judges of what constitutes health—but they are the best judges we have.) ▶ If the State runs both supply (money from general taxation) and demand (control of waiting lists etc), the rules of the market cannot operate and efficiency is hard to achieve—which is why some NHS trusts are being freed from central NHS control.

Controlling change—from on top: an example from maternity

- 1 Government sets up an expert group (mothers, midwives, ministers, obstetricians, and general practitioners—these are jokers in the pack, because they are simultaneously consumers, purchasers and providers).
- 2 Issuing of objectives and indicators of success—eg by 5 years:
 - Women should have a named midwife to ensure continuity of care.
 - Women should be able to choose their place of delivery. Aim to achieve the outcome that she believes is best for her baby and herself.
 - ≥75% of women should know the person who is to deliver them in labour.
 - Midwives should have direct access to some beds in all maternity units.
 - ≥30% of women should have a midwife as the lead professional.
- 3 The group's attractive-looking report is issued (using taxpayers' money) to all groups and personnel involved (except mothers).
- 4 Lack of finance is blamed when no improvements are detected at 5 yrs.

Anatomy of change Ideals (woman-centred care) → Specific policy objective (all women to have the chance to discuss their care) → Purchasers' action point (set up maternity services liaison committee with lay chairperson) → Providers' action point (provide link-workers, and advocacy schemes for women whose first language is not English). This type of activity may or may not lead to increased accountability and quality of services.

Referral statistics

There is great variability in individual GPs' referral statistics, which leads purveyors of government strategy to the error of saying 'Why is there a 4-fold difference in referral rates between GPs? Such variation is insupportable; some doctors must be referring too much'. An advance is made when this issue is reframed as: 'there is *information* contained in this variability, and this information can guide service development.'¹

Understanding the intricacies of purchasing health care depends on understanding referral patterns. If high-referring GPs refer unnecessarily, then the proportion of their referrals resulting in admission should be smaller than that of practices of similar size with low-referring GPs. Usually, this is not the case. Those with high-referral rates have high admission rates. How far does this relation hold? If I refer an ever-increasing number of my patients to a geriatric clinic, must a time come when admissions level off? The idea of a 'levelling-off effect' is important. If the consultant's actions are 'correct', and the GP's expectation as to the outcome of referral are uniform (probably never true) then when a levelling-off effect is observed, it may be true that the average referral rate is optimal, and that low-referrers are depriving patients, and high-referrers are wasting resources. In fact, levelling-off effects are rarely found—except in general surgery. What are we to conclude from most of the other specialties where no levelling-off effect is observed? Perhaps specialists admit a fixed proportion of patients referred to them. There is some evidence that this is true for ENT consultants and tonsillectomy. Another possibility is the Coulter–Seagroatt–McPherson hypothesis—that consultants have a threshold of severity for admission (eg a claudication distance of 50 metres) and even the majority of patients from the high-referrers fulfil this criterion. In this case (assuming the consultant is right), even the high-referrers are not referring enough. This may be true for all forms of angiography, for example. However, if the consultant is over-enthusiastic, and over-optimistic about the benefits of treatment, then the lower referrers are to be applauded for limiting the excesses of the consultant.

Overall, referral rates are no more variable than admission rates, even among populations with similar morbidities. The reason is probably that there is still a great deal of uncertainty underlying very many clinical decisions. We don't know who should have knee replacements, coronary angiography, cholecystectomy, aneurysm surgery, transplants, or grommets.

► There is no known relationship between high or low referral rates and quality of care. Here are 3 cautions in interpreting referrals:

- 1 Individual list size should not be used as a denominator, as it takes no account of differing workloads within a practice. Consultations per year would be a better denominator.
- 2 If doctors within the practice have special interests, these must be taken into account in comparing referral patterns.
- 3 Years of data are needed to compare referrals to rarely-used units.

Fitness to drive,^{UK} and other fitness issues

Ordinary UK driving licences issued by the DVLA (Driver & Vehicle Licensing Agency) are inscribed 'You are required by law to inform Drivers Medical Branch, DVLA, Swansea SA99 1AT at once if you have any disability (either physical or medical condition), which is, or may become likely to affect your fitness as a driver, unless you don't expect it to last >3 months.' It is the responsibility of the driver to inform the DVLA. It is the responsibility of their doctors to advise patients that medical conditions (and drugs) may affect their ability to drive and for which conditions patients should inform the DVLA. Drivers should also inform their insurance company of any condition disclosed to the DVLA. ► If in doubt, ask your defence union.

Heart conditions Uncomplicated MI: don't drive for 4 weeks. Angioplasty or pacemaker: don't drive for 1st post-op week. Angina: stop driving if symptoms occur at the wheel; DVLA need not be informed. Arrhythmias: driving may be OK once the cause has been found, and controlled for >1 month if the risk of ↓consciousness and motor power is sufficiently low. Syncope: OK to drive one week afterwards, if cause identified and treated. If no cause is found, stop for at least 6 months. TIA/stroke: stop for ≥1 month; no need to inform DVLA unless there is residual deficit for >1 month.

www.dvla.gov.uk/at_a_glance/content.htm

Diabetes All on oral hypoglycaemics or insulin must inform DVLA (in general, stop driving for 1 month after starting insulin, to get stable; drivers must demonstrate satisfactory control, and must recognize hypoglycaemia). Check that vision conforms to required standard (OPPOSITE). Avoid driving if hypoglycaemic risk ↑ (eg meal delay; or after excess exercise). Carry rapidly absorbed sugar in vehicle and stop, turn off ignition and eat it if any warning signs. A card should be carried to say which medications they are using to aid with resuscitation if needed. If an accident is due to hypoglycaemia a diabetic driver may be charged with driving under the influence of drugs.

CNS disorders Disabling giddiness, vertigo, and problems with movements preclude driving. DVLA need to know about unexplained blackouts, multiple sclerosis, Parkinson's (any 'freezing' or on-off effects), motor neurone disease, recurrent TIAs and strokes. In the latter the licence is usually withheld for 3 months depending on an examination by an independent doctor, and sometimes a driving test. Those with dementia should only drive if the condition is very mild (do not rely on armchair judgments: on-the-road trials are better). Encourage relatives to contact DVLA if a dementing relative should not be driving. GPs may desire to breach confidentiality (the GMC approves) and inform DVLA of demented or psychotic patients (tel. 01792 783686^{UK}). Many elderly drivers (~1 in 3) who die in accidents are found to have Alzheimer's.

For post-op driving in those with brain tumours, see below.*.

Epilepsy If a seizure while awake, he must not drive for 1yr. Attacks while asleep: stop driving for 1yr, unless the attack was while asleep and >3yrs ago and there have not been any awake fits since that asleep attack. In any event, the driving of a vehicle by such a person should not be likely to cause danger to the public. Provided a licence holder/applicant is able to satisfy the above, a 3yr licence will be normally issued. The 'till 70' licence may be restored if fit-free for 7yrs with medication if needed (if no other disqualifying condition).

Epileptic drug withdrawal risks a 40% seizure rate in year 1. Those wishing to withdraw from medication should cease driving from the beginning of withdrawal and not recommence until 6 months after treatment has ceased.

► For the most recent advice, always consult: www.dvla.gov.uk/at_a_glance/annex3.htm

Driving is prohibited in certain general categories:

- Severe mental disorder (including severe mental impairment).
- Severe behavioural disorders—or drug abuse/dependency.
- Alcohol dependency (including inability to refrain from drink driving).
- Psychotic medication taken in quantities that impair driving ability.

Vision Acuity (\pm spectacles) should be sufficient to read a 79.4mm-high number plate at 20.5 metres (\sim 6/10 on Snellen chart). Monocular vision is allowed only if the visual field is full. Binocular field of vision must be $>120^\circ$. Diplopia is not allowable unless *mild* and *eye-atch correctable*.

Drugs Driving, or being in charge of a vehicle when under the influence (including side-effect) of a drug is an offence under the Road Traffic Act 1988. Many drugs affect alertness and driving ability (check *Data-sheets*), and many are potentiated by alcohol, so warn patients not to drive until they are sure of side-effects, not to drink and drive, not to drive if feeling unwell, and never to drive within 48h of a general anaesthetic.

Old age DVLA says: 'progressive loss of memory, impairment in concentration and reaction time with possible loss of confidence, suggest consideration be given to cease driving.' This is too vague to help, as when reapplying for a licence (every 3yrs after 70yrs old) a driver simply signs to say 'no medical disability is present'. <http://bmj.com/cgi/content/full/324/7346/1154>

Fitness to fly: avoid hypobaric (high altitude) flights if...

- Climbing stairs causes troublesome dyspnoea (an easy screening test).
- Gas-filled dental caries (via putrifying bacteria): can cause severe odontalgia at altitude, and damage to the tooth.
- Within 48h after diving below 50 feet (the bends, p 802). Even at modest cabin altitudes death may occur. NB: barotrauma is worse on descent as the Eustachian tube is sucked flat by the low pressure in the middle ear, making the immediate equilibration of pressure more difficult.
- In uncontrolled cardiac failure. If O_2 supplementation is being used at sea-level, wean off before air travel, to help see if air travel is viable. All acute patients in this group should travel with enough supplementary O_2 to provide intermittent use eg at 2L/min.
- Confusional states and alcohol intoxication (synergistic with hypoxia).
- Pneumothorax; pneumomediastinum; or <10 days post-op to hollow organ.
- Neonates <3 days old, or women in the last 4 weeks of pregnancy (last 13 weeks if multiple pregnancy). See section on airlines, p 95.
- Anaemia (Hb <7.5 g/dL); GI bleeding; any recent tissue infarctions.
- Encourage good hydration and mobility; use aspirin & compression stockings on long-haul flights—p 81. NB: the list above is not exhaustive.

See International Air Transport Association advice. www.medinet.co.uk/crit.htm

Fitness to do sporting activities and cardiac rehabilitation

GPs often have to pronounce on this. Ensure that those involved know you do not have a crystal ball. Common sense, and attention to warm-up exercise is the key. If the patient is in an at-risk group, eg epileptics wanting to swim,⁶ or personal or family history of hypertrophic obstructive cardiomyopathy, and wanting to do heavy exercise, get help. medline:11790265
NB: in CCF, mild work with hand weights need not be banned. medline:11124492

See *Sports Medicine in Mims Companion*.

OK if well-controlled; go with a friend, only to lifeguard supervised pools. medline:8323342

Chronic fatigue syndrome (cfs)

cfs entails severe, disabling fatigue for >6 months, affecting physical and mental functioning, and present for more than 50% of the time. Myalgia, sleep disorders, and mood disturbance are common. www.bxt.co.uk/full.cfm?1058

Work-up of a patient with 'query chronic fatigue syndrome'

Exclude causes such as anaemia, TB, snoring, etc, with a history; physical examination; mental state (p 324), urinalysis, FBC, U&E, TSH, ESR, LFT, glucose \pm autoantibody screen, plasma creatine kinase, blood culture, and CXR.

Pursue abnormalities (eg ?TB/HIV if weight \downarrow ; ?depression if anhedonia, p336)

Does the pattern fit cfs?: persistent or relapsing fatigue, not relieved by rest, and leading to substantial reduction in previous levels of activity.

Are any exclusion criteria present?—psychosis (p 316), bipolar depression (p 354), dementia, anorexia nervosa (p 348).

Are 4 or more of the following present for >6 months?

- Impaired memory/concentration unrelated to drugs or alcohol use
- Unexplained muscle pain
- Polyarthralgia (but swelling suggests a joint diagnosis, p 675)
- Unrefreshing sleep
- Post-exertional malaise lasting over 24h
- Persisting sore throat not caused by glandular fever.
- Unexplained tender cervical or axillary nodes

If criteria met, call it cfs; if not fully met, call it 'idiopathic chronic fatigue'.

Co-morbid conditions are common, eg depression: consider diagnosing in a quantified way using formal diagnostic instruments (eg Beck inventory).

Treatment None is specific, and chronicity is common.

medline.9456063

Aim for a therapeutic alliance with your patient. Some therapists aim to prevent somatic fixation: the strongest predictor of a poor prognosis is a fixed belief that symptoms are due only to physical causes. Allow nonthreatening discussion about psychological issues, keeping an open mind on aetiology. Make it clear that psychological symptoms are not the same as malingering. 'Perhaps what starts as an illness may not be what keeps it going'. Psychological factors affect the outcome of many illnesses: why should this be different? Cautious increases in levels of activity are recommended (not quite the same as 'take more exercise'), to maintain muscle function, and improve confidence.

www.bmj.com/cgi/content/full/322/7283/38

Treat associated anxiety/depression (p 340). Address family and work problems. Talking with other patients sounds sensible, but there is a suggestion that this may prolong symptoms. Slow recovery is the norm.

Trials data. 3 randomized trials found that a graded exercise programme ('pacing') gave big improvement in fatigue and functioning.

medline.1065002

Relapses are triggered by too much exercise. Meta-analyses find cognitive therapy (p 370, via skilled therapists) can help. A trial found benefit from nicotinamide adenine dinucleotide (NADH) 10mg/24h (8/26 improved, compared with 2/26 in on placebo; NADH helps generate ATP which may be depleted in cfs).

www.bxt.co.uk/sum.cfm?1042

When to get help ● Children with cfs ● Unresponsive to the above measures ● History of travel abroad ● CNS signs ● Walking difficulty ● Fevers ● Suicidal.

Managing obesity

Obesity (BMI >30) shortens life, aggravates diabetes, gout, osteoarthritis, heartburn, BP↑, lipid↑, sleep apnoea, and DVT. Weight loss in the obese diminishes BP and osteoarthritis, and restores life expectancy to normal.

The hypothalamus with its 40-or-so neurotransmitters regulates appetite and energy balance, by integrating hormonal, neural, and peripheral messages (concerning gut distension). Neuropeptide γ is a transmitter stimulating feeding, and suppressing the sympathetic nervous system (\therefore ↓energy expenditure). Other transmitters increase release of serotonin and noradrenaline from nerve terminals, and act as mediators of satiety, as does peripherally produced cholecystikinin. Leptin is thought to be a bloodborne signal from the adipose tissue that informs the brain about the size of the fat mass. Animal models show that highly palatable foods lead to obesity, limited by 'futile thermogenesis', a metabolic cycling achieved by genetically controlled uncoupling oxidative phosphorylation in mitochondria. Genes for uncoupling proteins have been cloned, and may lead to novel treatments for obesity.

Desirable weights (see opposite). A guide to excessive intra-abdominal fat (correlates best with obesity's problems) is a waist >94cm in men. ~1/3 of a GP's list is overweight by this criterion: *the richer a society, the more prevalent is obesity*. Despite 33,000 publications on obesity and spending \$33 billion/yr (USA), there are no good, well-tried nonsurgical nondrug techniques for long-term maintained weight loss.

Prevention by education can work, eg in school, is one way forward.

Fat & fat free mass (FFM) Excess weight is ~75% fat and 25% FFM, (water, protein & glycogen—energy value 1000kcal/kg). With very low calorie diets (<330kcal/24h) FFM loss is too high. Exercise accounts for 20% of energy expenditure (so contributes much less to weight loss than diet). Simple lifestyle changes (eg 2 brisk walks/day) are more likely than supervised exercise to produce long-term benefit; but don't rely on this to ↓weight. EBM 7 73

Very Low Calorie Diets (VLCDS) give 400kcal/24h with 40g protein/24h for women, 500kcal/24h and 50g protein/24h for men, and women >173cm. 2 pints of skimmed milk with bran for bulk and unlimited calorie-free drinks gives the former, 3 pints the latter. All should take 200mg ferrous sulfate and one multivitamin tablet/24h. This is cheaper than commercial VLCDS. As excess fat free mass is lost, use for >4 weeks needs medical supervision.

Drugs for which there is randomized evidence of effectiveness

▶ No drug is a panacea capable of making fat people thin effortlessly.

Clistor may be used for ≤2yrs in those with BMI >28kg/m² + associated risk factors who have shown weight↓ on a diet (which must be continued). CI: malabsorption. SE: (little is absorbed) oily faecal incontinence/spotting, flatus, urgency. This is reducible by co-therapy with natural fibres (psyllium hydrophilic mucilloid). medline.11443512 Dose: eg 120mg with a meal (up to 3/day, =£560/yr); stop after 12 weeks if weight loss <5%. medline.11399238

Serotonin reuptake inhibitors inhibit serotonin & noradrenaline reuptake, promoting weight loss by enhancing postingestive satiety, and increasing calorie use. It's approved by NICE in those aged 18–65 if serious attempts at weight loss have failed, and BMI is >27 with associated problems (eg DM)—or if BMI >30 and no associated problems. SE: CCF/IHD; BP↑. SI: pulse↑; BP↑; dry mouth. 2001 medline.11407998

NB Amphetamine-like appetite suppressants are no longer recommended.

Surgery is reserved for the morbidly obese (BMI >40; or >35 with associated cardiovascular risk factors). Gastric bypass and gastroplasty result in sustained weight loss. SE: dumping (OHCM p 514) and vitamin deficiency.

Men	Height		Acceptable weight range						Overweight			Obese		
	m	ft in	kg	st	lb	–	st	lb	kg	st	lb	kg	st	lb
58	5	2	44–64	6	13	–	10	1	70	11	0	77	12	2
60	5	3	44–65	6	13	–	10	3	72	11	5	78	12	4
62	5	4	46–66	7	3	–	10	6	73	11	7	79	12	6
64	5	4	47–67	7	6	–	10	8	74	11	9	80	12	8
66	5	5	48–69	7	8	–	10	12	76	12	0	83	13	1
68	5	6	49–71	7	10	–	11	3	78	12	4	85	13	5
70	5	7	51–73	8	0	–	11	7	80	12	8	88	13	12
72	5	8	52–74	8	3	–	11	9	81	12	11	89	14	0
74	5	9	53–75	8	5	–	11	11	83	13	1	90	14	2
76	5	9	54–77	8	7	–	12	2	85	13	5	92	14	7
78	5	10	55–79	8	9	–	12	6	87	13	10	95	14	13
80	5	11	58–80	9	2	–	12	8	88	13	12	96	15	2
82	6	0	59–82	9	4	–	12	13	90	14	2	98	15	6
84	6	0	60–84	9	6	–	13	3	92	14	7	101	15	13
86	6	1	62–86	9	11	–	13	8	95	14	13	103	16	3
88	6	2	64–88	10	1	–	13	12	97	15	4	106	16	10
90	6	3	66–90	10	6	–	14	2	99	15	8	108	17	0
92	6	3	68–93	10	10	–	14	9	102	16	1	112	17	9

Women	Height		Acceptable weight range						Overweight			Obese			
	m	ft	in	kg	st	lb	–	st	lb	kg	st	lb	kg	st	lb
48	4	10		37–53	5	12	–	8	5	58	9	2	64	10	1
50	4	11		37–54	5	12	–	8	7	59	9	4	65	10	3
52	5	0		38–55	6	0	–	8	9	61	9	8	66	10	6
54	5	1		39–57	6	2	–	9	0	63	9	13	68	10	10
56	5	1		39–58	6	2	–	9	2	64	10	1	70	11	0
58	5	2		40–58	6	4	–	9	2	64	10	1	70	11	0
60	5	3		41–59	6	6	–	9	4	65	10	3	71	11	3
62	5	4		42–61	6	9	–	9	8	67	10	8	73	11	7
64	5	5		43–62	6	11	–	9	11	68	10	10	74	11	9
66	5	5		44–64	6	13	–	10	1	70	11	0	77	12	2
68	5	6		45–65	7	11	–	10	3	72	11	5	78	12	4
70	5	7		45–66	7	11	–	10	6	73	11	7	79	12	6
72	5	8		46–67	7	3	–	10	8	74	11	9	80	12	8
74	5	9		48–69	7	8	–	10	12	76	12	0	83	13	1
76	5	9½		49–70	7	10	–	11	0	77	12	2	84	13	3
78	5	10		51–72	8	0	–	11	5	79	12	6	86	13	8
80	5	11		52–72	8	3	–	11	9	81	12	11	89	14	0

Desirable weight relates to body mass index (BMI), W/H^2 . W =weight (kg); H =height (metres); mortality is least if 20–25. **For** a given weight, **trunk** (central) obesity is worse than **devolved** obesity (perhaps because mental fat causes insulin resistance via a molecule known as resistin).¹

Dietary approaches

Ask patients to keep a diary of foods eaten.

Aim for realistic loss (eg <1 kg/week for 1000kcal per day diet).

Regular (weekly) weighing to watch trends.

Consider weighing food to calculate calorie intake to prevent 'cheating'.

Groups such as Weight Watchers can be very useful motivating forces.

Aim to re-educate eating habits to maintain reduced weight.

Ensure that the diet is nutritious. * See the *Healthy Diet*, *OHCM* p 194.

even (Mayo Clinic) questions for weight-loss programmes to answer

Is there proof that it works (ask for evidence of long-term results)?

Any qualified dietitian or specialist in behaviour modification employed?

Is the recommended intake nutritionally balanced?

Does the patient have to buy special products?

Will the patient receive advice on starting safe, moderate exercise?

How will the programme reward and monitor progress?

Is there support and follow-up for after the formal programme ends?

What to do for the best

Doctors are called on to make decisions about every patient they meet: few are curable at once, so making a plan for what to do for the best is the secret of success at the bedside. The aim here is to explain this secret, to enable you to flourish in the clinical world, and to keep you out of lawyers' offices.

Let us look at the steps of the history, physical, or mental examination, and investigations.

By the end of taking the **history**, you need to have acquired 3 things:

- 1 Rapport with the patient
- 2 A diagnosis or differential diagnosis
- 3 The placement of the diagnosis in the context of the patient's life

Rapport Consultations are shorter when rapport is good. medline:10944650

The patient is confident that he or she is getting the full attention of the doctor, and these patients are more understanding, and more forgiving when things go wrong. Doctors are far from infallible: so we need to have confidence that the patient will feel able to come back if things are not right, tell us what has happened, agree on an adjustment of the treatment, and, by giving feedback, improve our clinical acumen.

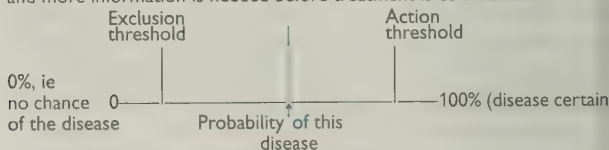
Diagnosis Studies have shown that skilled physicians have made a provisional diagnosis soon after the consultation starts, and they spend the rest of the history in confirming or excluding it. What happens if you are not skilled, and you have no hint as to the diagnosis? You need to get more information:

- Pursue the main symptom: 'tell me more about the headache. . .'
- Elicit other symptoms—eg change of weight or appetite, fevers, fatigue unexplained lumps, itching, jaundice, or anything else odd?
- Get help from a colleague or even a diagnostic system—eg Mentor, p 446.
- Check you still have rapport with the patient. Are you searching for a physical diagnosis when a psychological diagnosis would be more appropriate? Here you might ask questions such as 'How is your mood?' 'What would your wife or partner say is wrong?' 'Would they say you are depressed?' 'What would have to change for you to feel better?'

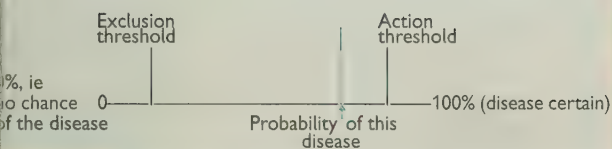
► Do not proceed to the physical examination until you have a working diagnosis: the answer is rarely found there (<10%).

Placing the diagnosis in the context of the patient's life If you do not do this, you will not know what will count as a cure, and, more specifically, different patients need different treatments—see p 1. Some factors to focus on might be: the motivation of the patient to get better ('I've got to get my knee better so that I stay strong enough to lift my wife onto the car mode'); their general health; social situation; drugs (not forgetting nicotine and alcohol); is help available at home; work (yes/no; type)?

At the end of the history, occasionally there is enough information to start treatment. Usually you may be only, say, 70% sure of the diagnosis and more information is needed before treatment is commenced.



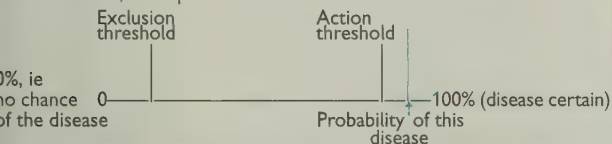
It is time for the physical **examination**. This aims to gain evidence to confirm or exclude the hypothesis, to define the extent of some process, or to assess the progress of known disease. At each step, ask 'what do I need to know?' Following the examination the diagram may look like this:



Investigations If the action threshold has not been crossed, further information is needed. Action thresholds vary from doctor to doctor, and from disease to disease. When the treatment is dangerous, the action threshold will be high (eg leukaemia). In self-limiting illnesses, eg pharyngitis, the action threshold will be lower. Note that 'action' may be that, in agreement with such a patient, only symptomatic treatment is needed, and future episodes could be managed without medical input.

Similarly, it may be important to move the probability of a serious but unlikely disease beyond the exclusion threshold.

Once the probability of a disease passes the action threshold, treatment can commence, if the patient wishes.



Supposing neither the action threshold, nor the exclusion threshold is exceeded, then more information is needed, eg from pathology, imaging, or from the passage of time. Time itself is an investigation: it may reveal sinister causes or the benign nature of the disease. To use time this way, you need to be reasonably sure that immediate treatment is not required.

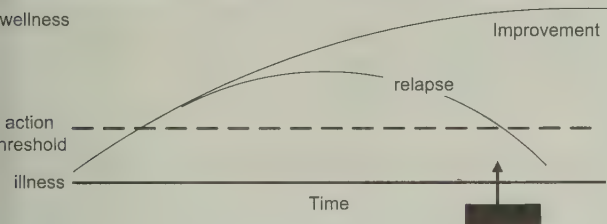
If there is still not enough certainty to initiate management, get further information, eg from books, computers, colleagues, further tests—or you may feel it appropriate to refer the patient at this stage. Or go round the process again, starting with the history—from a different view point.

Once above the action threshold, it is time to decide what to do for the best. This is a decision shared by the doctor and the patient. It entails informed consent and consideration of:

medline:1454004

- The probability of the diagnosis
- The likelihood of the different possible outcomes
- The costs and side-effects of treatment
- The hope and values of those affected, particularly the patient
- What is possible, considering the skills, resources, and time available

Finally, tell your patient how they will know if they are on the path to improvement or relapse, and if so, at what point to seek help (critical action threshold, below; record this in the notes)—eg if your peak flow falls by 40%, start this prescription for prednisolone, and come and see me.



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Sources: 1 Eye sites: www.neri.harvard.edu/ (tel. +44 (0)20 725 33411)

www.bjophthalmol.com/ Royal National Institute of the Blind.

www.massachusetts-eye.com/ Royal College site with links to the journal Eye.

www.eri.harvard.edu/ The Massachusetts Eye and Ear Infirmary

(The Schepens Eye Institute)

Journals www.bjophthalmol.com/; www.bmj.com

We especially thank Mr P Muthusami FRCS & Mr R Williams FRCS for their help.

We do not see the human eye as a receiver, it appears not to let anything in, but to send something out. The ear receives; the eye looks. (It casts glances, it flashes, radiates, gleams.) One can terrify with one's eyes, not with one's ear or nose. When you see the eye, you see something going out from it. You see the look in the eye. If you only shake free from your physiological prejudices, you will find nothing queer about the fact that the glance of the eye can be seen too. For I also say that I see the look that you cast at someone else. *Vertigenste*: (Zettile: 222-3).

- Accommodation** The active changing of lens shape to focus near objects.
- Acuity** A measure of how well the eye sees a small or distant object.
- Anisometropia** Reduced acuity which is not from an anatomic optic defect.
- Anisometropia** Test chart of intersecting lines used for screening for macular disease. If present, lines appear wavy and squares distorted.
- Anisocoria** Unequal pupil size.
- Anisometropia** Having different refractive errors in each eye.
- Anophthalmos** The state of having no lens (eg removed because of cataract).
- Blepharitis** Inflamed lids.
- Canthus** The medial or lateral angle made by the open lids.
- Chemosis** Oedema of the conjunctiva.
- Choroid** Vascular coat between the retina and the outer scleral coat.
- Ciliary body** Portion of uvea (uveal tract) between iris & choroid, containing ciliary processes & ciliary muscle (for accommodation).
- Conjunctiva** Mucous membrane on anterior sclera & posterior lid aspect.
- Cycloplegia** Ciliary muscle paralysis preventing accommodation.
- Dacryocystitis** Inflammation of the lacrimal sac.
- Dioptres** Units for measuring refractive power of lenses.
- Ectropion** The lids evert (especially lower lid).
- Entropion** The lids invert (so that the lashes may irritate the eyeball).
- Epiphora** Passive overflow of tears on to the cheek.
- Conjunctiva** Where bulbar (scleral) and palpebral (lid) conjunctivae meet.
- Fovea** The tiny, vital, cone-rich area of retina capable of 6/6 vision.
- Macula** That part of retina normally visible through the ophthalmoscope.
- Keratoma** The cornea is shaped like a cone. See p 520.
- Keratomalacia** The cornea is softened.
- Limbus** The annular border between clear cornea and opaque sclera.
- Machula** Rim of avascular retina surrounding the fovea.
- Miotic** An agent causing pupil constriction (eg pilocarpine).
- Mydriatic** An agent causing pupil dilatation (eg tropicamide).
- Near point** Where the eye is looking when maximally accommodated.
- Optic neuritis** Inflammation of the optic nerve head.
- Optic disc** The cup like depression in the centre of the optic disc (p 503).
- Optic disc** That part of optic nerve seen ophthalmoscopically in the fundus.
- Presbyopia** Age-related reduced near acuity from failing accommodation.
- Pterygium** Wing-shaped degenerative conjunctival condition (p 480).
- Ptosis** Drooping lids.
- Refraction** Ray deviation on passing through media of different density; or determining refractive errors, and correcting them with lenses.
- Retinal detachment** The sensory retina separates from the pigmented epithelial layer of retina.
- Sclera** The whites of the eyes starting from the corneal perimeter.
- Scleroma** A defect causing a part of the field of view to go missing.
- Slit lamp** A device which illuminates and magnifies structures in the eye.
- For slit lamp images of the above conditions, see www.txt.co.uk/full.cfm?1088
- Squint** (squint) Eyes deviate (they are not looking at the same thing).
- Tarsorrhaphy** A surgical procedure for uniting upper and lower lids.
- Tonometer** A device for measuring intraocular pressure.
- Uvea** Iris, ciliary body and choroid.
- Vitreous** Jelly-like substance filling the globe behind the lens.
- Vitrectomy** Surgical removal of the vitreous.

Examination of the eye

The history indicates the part of the examination to focus on. Asking about trauma, symptoms of pain vs irritation, loss of vision, and pattern and speed of onset. Examination assesses the retina and optic nerve (nerve of vision), tests acuity, visual fields and colour vision (p 517).

Acuity is a measure of central (macular) vision; always test it carefully as any loss may be serious. Record it accurately, especially in a patient with eye injury. Examine the right eye first. Sit the patient 6 metres from the Snellen chart (p 479); to get 6 metres in a 4 metre room, place the chart just above the patient, pointing towards a mirror 3 metres away. Obscure the left eye with a lollipop-shaped 'eye paddle' or card, not pressed against the eye—simply enough to occlude the visual access (more reliable than peepable-through fingers or a slipping palm). Ask the patient to read it from the top using the right eye, then the left. Use glasses if worn. The last line completed accurately indicates the acuity for distant vision.

The chart is designed so that the top line can be read by someone with normal vision at 60 metres, the next at 36 metres, the next at 24, the next at 18, the next at 12, the next at 9, and the next at 6 metres. Acuity is recorded as 6/60, 6/36, 6/24, 6/18, 6/12, 6/9, 6/6 to indicate the last line accurately read (6/6 vision is normal). For acuities of worse than 6/60 the patient can be brought forward to 5, 4, 3, 2 and 1 metre from the chart to read the top line. If he can read it then acuity is expressed as that distance, eg 5/60, 4/60, 3/60, 2/60 or 1/60. If the vision is below 1/60 ask the patient to count your fingers at ½ metre distance. This is recorded as cf (count fingers). If they cannot count your fingers move your hand in front of the eye at ¼ metre distance. If the patient can appreciate that your hand moves, record hm (hand movement). If the patient cannot appreciate hand movement, dim the light in the examination room and shine a torch light into the eye. If the patient perceives light, record pl. If there is no perception, record 'no pl' (the eye is blind). NB: in practice, if nothing on the Snellen chart can be read, it is common to go straight to finger counting.

If the patient sees less than 6/6 with or without glasses, examine again with a pin-hole in front of the eye: a narrow beam removes the need for focus. In simple refractive errors, acuity will improve through the pin-hole. This is an important test as it eliminates ocular pathology as a cause of reduced acuity. (Make a pin-hole with a 22G needle in a 10 × 10cm opaque card; check that you can see through the hole before giving it to the patient.)

If patients >40 years old complain of near-vision blurring, the cause may be presbyopia (p 490). Test near vision using a near vision testing card (p 478). If the patient can read N5 at 30cm, near vision is normal.

Visual field This is the area that can be seen with both eyes without shifting gaze. The uni-ocular field is smaller than the binocular field. Questions to address: does the defect affect one eye or both? Are there any clear boundaries to the defect? Does the boundary lie in the vertical or horizontal meridians? Is acuity affected? (see front of tests p 492).

External examination: Is it symmetrical? Normally retraction on upward gaze (abnormal in dysthyroid disease). Note ptosis (p 480), spasm, inflammation or swellings (p 480). Look for inflammation (if circumcorneal, suspect anterior uveitis; injection of the bulbar, fornix, and the tarsal surfaces suggests conjunctivitis; focal injection adjacent to the cornea 'points' to a problem on the cornea). Is there discharge, follicles, or cobblestone patterns on tarsal conjunctiva of upper lid, or any subconjunctival haemorrhage (p 496)? Examine with a torch for opacity, abrasion, ulcer (the latter two stain green with 1% fluorescein), or oedema.

Anterior chamber: It is filled with clear aqueous humor but can be cloudy in anterior uveitis (p 494), may have sterile pus (hypopyon) with corneal ulcer, or blood (hyphaema) after injury.

Pupils should be equal, reacting to light and accommodation (PERLA). They are small and irregular in anterior uveitis, dilated and fixed in acute glaucoma (p 494). For other pupillary abnormalities see p 488.

Lenses: When the lens is normal the pupil appears black. With mature cataract it may appear white.

Extraocular movements It is vital to examine these in those with diplopia. Ask the patient to watch a pencil move diagonally: up left; up right; down left; down right; horizontally left; horizontally right. Ask which movement provokes most diplopia, and when looking in that direction, block each eye in turn and ask which one sees the *outer* image: that is the eye which is malfunctioning. NB: avoid extremes of movement as inability to maintain fixation stimulates nystagmus. For eye movements and squint see p 486.

Ophthalmoscopy This helps detect pathology in the lens, vitreous, and retina. Start with high + numbers (often marked in colour on the dial). To examine the lens and the vitreous focus the beam of the ophthalmoscope at the pupil at ~1 metre from the eye. In the normal eye there is a red glow from the choroid (the red reflex). Any lens opacity (cataract) will be seen as a black pattern obstructing the red reflex. Blood or loose floaters in the vitreous are seen as black floaters. To determine the position of the opacity in the eye, move your head in the horizontal plane from side to side, like a cobra waiting to strike. The opacity will either move in the same direction as you (so lies behind the lens), in the opposite direction (so lies in front of the lens) or will not move at all (so *in* the lens).

Red reflexes are absent with dense cataract and intraocular haemorrhage. When the retina is in focus, ask the patient to look at the Snellen chart or any other object right ahead at the end of the room. This will bring the optic disc into the view. Examine it carefully; it should have precise boundaries and a central cup (p 502). Cupping means an excavated appearance (seen in glaucoma). A pale disc suggests optic atrophy (eg chronic glaucoma, multiple sclerosis, *et al.*, p 500). Examine radiating vessels and macula (ask the patient to look at the light).

Tips to successful ophthalmoscopy: Ensure the batteries are fully charged. Darken the room; remove spectacles; dial up a lens to correct the resulting refractive error (– lenses correct myopia, + correct hypermetropia). If very myopic, try examining with spectacles on (discs appear small). If you find ophthalmoscopy difficult using your non-dominant eye, try using your dominant eye for examining *both* fundi—while standing behind the seated patient, whose neck is fully extended.

Always check the lens for opacities before trying to examine the fundus. Get *close enough* to the patient, even if one of you has had garlic for lunch. Consider using a short-acting mydriatic to dilate the pupil (see p 518). Remember that most retinal tears are peripheral and are difficult to see. It isn't possible to see the margin of the retina with an ophthalmoscope.

Indirect ophthalmoscopes have much wider fields of view. The skull makes contact with its eyecup. Good views are possible even via quite constricted pupils. Magnification and ↑field width is achieved by focusing a beam on the cornea, which is then fanned out. www.one-stopmedical.com 01494 722480

Slit lamp examination This instrument has a bright light source and a horizontally mounted microscope to examine the structures of the living eye. The light source can be converted to a slit (hence the name). Gonioscopic attachments allow intraocular pressure measurement.

N. 48

He moved

N. 36

forward a few

N. 24

fine chattering gems.

N. 18

He knew exactly who would

N. 14

now sneeze calmly through an open door. Had there been another year

N. 12

of peace the battalion would have made a floating system of perpetual drainage.

N. 10

A silent fall of immense snow came near oily remains of the purple-blue supper on the table.

N. 8

We drove on in our old sunless walnut. Presently classical eggs ticked in the new afternoon shadows.

N. 6

We were instructed by my cousin Jasper not to exercise by country house visiting unless accompanied by thirteen geese or gangsters.

N. 5

The modern American did not prevail over the pair of redundant bronze puppies
The worn-out principle is a bad omen which I am never glad to ransom on purpose

H
A L
T N C
O L H A
E C T N O
C L O H N A
A E N L O N C T



H



T



O Y



H U V

A T Y M

X O W U H



Y U V T X O

Reading types are read at 30cm (with reading glasses if used)

Lines on the snellen chart are as follows: (with distance glasses)

6 Able to read at 6m what can normally be read at 6m, ie '6/6'

9 Able to read at 6m what can normally be read at 9m, ie '6/9'

12 Able to read at 6m what can normally be read at 12m, ie '6/12'

18 Able to read at 6m what can normally be read at 18m, ie '6/18'

24 Able to read at 6m what can normally be read at 24m, ie '6/24'

36 Able to read at 6m what can normally be read at 36m, ie '6/36'

60 Able to read at 6m what can normally be read at 60m, ie '6/60'

Counts fingers; counts fingers held $\frac{1}{2}$ m distance, ie 'CF'

Hand movement; perceives hand moving $\frac{1}{4}$ m distance, ie 'HM'

Perceives light; can see a torchlight when shone into eye, ie 'PL'

No light perception, abbreviated to 'no PL', ie blind.

The external eye

Entropion (Inturning of the lids, especially lower). Usually due to degenerative changes in the fascial attachments of the lower lids and their musculature. It is rare if <40yrs old. The inturned eyelashes irritate the cornea. Taping the (lower) eyelids to the cheek gives temporary relief; more lasting relief needs surgery. medline:11056776

Ectropion Lower lid eversion causes eye irritation, watering (drainage punctum malaligned) ± exposure keratitis. **Associations** Old age; facial palsy. Plastic surgery may correct the deformity. If facial palsy is the cause, consider eg a modified temporalis muscle transfer. medline:10597819

Upper lid position may result from the globe's hypotropic position or excess tissue in the lid—**pseudoptosis**, or intrinsic levator weakness,—**true ptosis** from: • Congenital (absent nerve to the levator muscle; poorly developed levator); • Mechanical (oedema, xanthelasma or tumour of the upper lids); • Myogenic (muscular dystrophy, myasthenia); • CNS (III nerve palsy, p 486; Horner's, p 488). Congenital ptosis is corrected surgically early if the pupils are covered or if it is unilateral (risk of amblyopia ex anopsia).

Lagophthalmos This is difficulty in lid closure over the eyeball. **Causes** Exophthalmos; mechanical impairment of lid movements (eg injury or lid burns); leprosy; paralysed orbicularis oculi giving sagging lower lid; infective keratitis. medline:10832688 Corneal ulceration and keratitis may follow. Lubricate eyes with liquid paraffin ointment. If corneal ulceration develops, temporary tarsorrhaphy (stitching lids together) may be needed.

Styes The word stye is used more by patients than by ophthalmologists for referring to inflammatory eyelid swellings. **Hordeolum externum** is an abscess or infection, usually staphylococcal, in a lash follicle; these may also involve the glands of Moll (sweat glands) and of Zeis (sebum-producing glands attached directly to the lash follicle). They 'point' outwards and may cause extensive inflammation of the eyelids. Treatment is with local antibiotics—eg fusidic acid. (The word 'stye' implies infection: if this is not present, the term **marginal cyst**—of Zeis or Moll—may be used.) Less common is the **hordeolum internum**, an abscess of the Meibomian glands (**hordeolum** is Latin for *barley*corn). These 'point' inwards, opening on to conjunctiva, cause less local reaction but leave a residual swelling called a **chalazion** or a **Meibomian cyst** (tarsal cyst) when they subside. Vision may be ↓ if corneal flattening occurs (rare). medline:9269001 Treatment for residual swellings is incision & curettage (+ eg fusidic acid applied for some days pre-op). Lidocaine-prilocaine cream can obviate need for GA. medline:8782255

Blepharitis (Lid inflammation, eg from staphs, seborrhoeic dermatitis, or rosacea). Eyes have 'burning' itching red margins, with scales on the lashes. **Treatment** Cleaning crusts off the lashes is essential (use cotton-wool buds) ± Tears Naturale®, fusidic acid, or steroid drops (or creams). In children with blepharokeratitis, consider oral erythromycin too. medline:11124676

Pinguecula Degenerative yellow nodules on the conjunctiva either side of the cornea (typically nasal side). **Treatment** Adult male. **Associations** ↑ hair & skin pigment; sun-related skin damage. medline:9685008 If they become inflamed (pingueculitis)—they may respond to topical steroids. If it encroaches on to the cornea, as it may in dusty, wind-blown life-styles, the word **pterygium** is used (surgery may be needed). www.bxt.co.uk/full.cfm?1088

Dendritic ulcer (*Herpes simplex* corneal ulcer) **Signs** Photophobia & epiphoria (watering). If steroid drops are used, corneal invasion and scarring may occur, risking blindness. **Diagnosis** 1% fluorescein drops stain the lesion. **Rx** Aciclovir 3% eye ointment 5 × daily for ≥3 days after complete healing. Get expert help if aciclovir resistance seems possible. medline:11190039 & 7600820

Tears and lacrimation

The lacrimal glands are on the superior temporal side of the orbits. The tear film excreted over the eye drains via the lacrimal puncta (found at the medial side of the upper and lower eyelid) through the lacrimal sac, lacrimal duct and inferior meatus (just lateral to the inferior turbinate) into the nasal passages. Dry eyes may be due to insufficient tear secretion, and watering eyes may be due to blockage of the drainage system.

Acute dacryocystitis This is acute inflammation of the tear sac which is located medial to the medial canthus. This may spread to surrounding tissues and result in systemic upset. Immediate antibiotic therapy may resolve the infection. Failure leads to local abscess formation.

Nasolacrimal duct non-canalization The nasolacrimal duct may not be canalized at birth and may not open fully until 3 months old. Tear sacs tend to get infected and a sticky discharge is produced. Ask an ophthalmologist to teach the mother to massage the sac to empty the contents four times daily and then apply antibiotic (eg gentamicin) eye drops. Should this fail after several months, probing of the duct under anaesthesia is an option.

Chronic dacryocystitis This typically occurs in the middle-aged and elderly. There is lacrimal sac distension, discharge of mucopus into the eye \pm nasolacrimal duct block. Syringing the lacrimal drainage system is done early to clear the system; it may need repeating. If the nasolacrimal duct is permanently blocked, dacryocystorhinostomy (to establish communication between the lacrimal sac and the nasal cavity) is needed. Squamous cell cancer of the lacrimal drainage system: do radiographic examination (eg CT, MRI) of all masses arising in the medial canthus. medline:9825006

Dacryoadenitis Lacrimal gland inflammation causes pain and swelling on the temporal side of the upper eyelid, which may become S-shaped. Causes: Viral (mumps, measles, influenza) or gonococcal. Chronic swelling can occur in sarcoid, TB, lymphatic leukaemia, or lymphosarcoma. www.fpnotebook.com/EYE53.htm

Tear production The volume of tears normally in the eye is $6\mu\text{L}$, the turnover rate being $1.2\mu\text{L}/\text{min}$. Tears are similar in electrolyte concentration to plasma, but rich in proteins, especially IgA. They also contain lysozyme and β -lysin which have antibacterial properties.

Dry eye syndrome This may be due to \downarrow production of tears by the lacrimal glands in old age, or, less commonly, in: Sjögren's syndrome associated with connective tissue disorders (especially RA); mumps; sarcoidosis; amyloid; lymphoma; leukaemia; haemochromatosis. Other causes: \downarrow (post-exposure keratitis); or \downarrow (avitaminosis A, Stevens-Johnson syndrome, pemphigoid, chemical burns). Schirmer's test (strip of filter paper put overlapping lower lid; tears should soak $>15\text{mm}$ in 5min) reveals insufficient production. Artificial tears may be used for symptomatic relief.

Excess lacrimation Emotion (joy or sorrow),* corneal abrasions or foreign body, conjunctivitis, iritis, acute glaucoma.

Ectropion ie normal volume, but not normally drained to the inferior meatus of the nose). Ectropion, entropion, blockage of drainage system.

Dacryocystorhinostomy (eg endoscopic) for nasolacrimal duct obstruction; surgery for other causes. medline:11020191

*Some sceptics argue that tears of joy don't exist. Sandor Feldman claims they are really tears of anticipated loss (T Lutz, 1999, *Crying, the Natural and Cultural History of Tears*, ISBN 0393047563 & K Clanan *Lancet* 2000 **355** 663 www.txt.co.uk, full cfm 0904). In support of this, the 'fact' is cited that children do not cry with tears of joy. To this we simply reply that psychoanalysts cannot have much joy in their families—or else they'd have observed infant tears of joy at family reunions.

Orbital swellings

Lesions in the bony orbit typically present with proptosis (ie exophthalmos), whatever the pathological origin. Proptosis (protrusion of orbital contents) is a cardinal sign of intraorbital problems. If pressure is eccentric within the orbit there will be deviation of the eyeball \pm diplopia. Orbital pain usually arises from neighbouring structures (eg sinusitis).

Orbital cellulitis This usually results from spread of infection from the paranasal air sinuses (or eyelid, dental injury/infection, or external ocular infection). *Typical patient* A child with inflammation in the orbit, with fever, lid swelling, and \downarrow eye mobility. *Causes* Staphs, *Strep pneumoniae*, *Strep pyogenes*, *Strep milleri*. Admit for prompt CT, antibiotics (cefuroxime 1.5g/8h iv; child 20mg/kg/8h) \pm surgery—to prevent extension to the meninges and cavernous sinus. Blindness is a risk from pressure on the optic nerve or thrombosis of its vessels. medline:11148155 &: 10804112

Carotico-cavernous fistula is usually due to carotid aneurysm rupture with reflux of blood into the cavernous sinus. *Causes*: Spontaneous; trauma; post-septorhinoplasty. There is engorgement of the eye vessels + lid & conjunctival oedema. Exophthalmos may be pulsatile, with a loud bruit over the eye \pm tinnitus. Symptoms may subside spontaneously. Arterial ligation or embolization may be needed. medline:7110490 &: 8540139 &: 7825255

Orbital tumours Primary neoplasms are rare (angiomas, dermoids, meningiomas, or gliomas of the optic nerve). Secondary tumours are more common. Reticuloses can form orbital deposits (examine liver, spleen, nodes; do FBC). In children unilateral proptosis may be the first sign of a neuroblastoma. Nasopharyngeal tumours occasionally invade the orbit, as may mucocoeles and pyocoeles of the ethmoid and frontal sinuses. CT scan pictures give a clear representation of the orbit.

Hyperthyroidism may cause exophthalmos (*OHCM* p 295).

Ophthalmic shingles

This is zoster of the 1st (ophthalmic) branch of the trigeminal nerve and accounts for 20% of all shingles. Only thoracic nerves are more affected (55%). Pain, tingling or numbness around the eye may precede a blistering rash which is accompanied by much inflammation. In 50% of those with ophthalmic shingles the eye itself is affected with 40–77% having corneal signs and 50–60% iritis. Nose-tip involvement—Hutchinson's sign—means involvement of the nasociliary branch of the trigeminal nerve which also supplies the globe and makes it highly likely that the eye will be affected. The eye can be seriously involved with little rash elsewhere. Beware dissemination if immunocompromised. medline:9279053

Presentation:

- | | | |
|--------------------------------|-----------------|---------------------------|
| ● Mucopurulent conjunctivitis | ● Episcleritis | ● Iritis (\pm atrophy) |
| ● Limbal lesions ¹ | ● Visual loss | ● Pupillary distortion |
| ● Preauricular node tenderness | ● V nerve palsy | ● Optic atrophy |
| ● Scleritis | ● Keratitis | |

Treatment Aciclovir 800mg 5 times daily po for 7 days to \downarrow viral shedding, accelerate healing, and \downarrow incidence of new lesions. Start within 4 days of onset. It is wise for all with ophthalmic shingles to see a specialist if the nose-tip is involved or the eye turns red within 3 days to exclude anterior uveitis with a slit lamp. Prolonged steroid eyedrops may be needed.

Retinoblastoma

This is the most common primary intraocular tumour in children.

Incidence 1 in 15,000 live births.

Signs Strabismus and leukocoria (ie a white pupil). Always suspect retinoblastoma when the red reflex is absent (the mother may come with a photo showing only one eye reddened during flash photography); NB: multiple tumours may be present. medline:10101941

Inheritance Hereditary retinoblastoma is different from the non-hereditary type: there is a mutation of the Rb gene located at 13q14. Inheritance is autosomal dominant with 80% penetrance. The RB gene is present in everyone, and is normally a suppressor gene or anti-oncogene. Those with hereditary retinoblastomas typically have one altered allele in every cell. If a developing retinal cell undergoes mutation in the other allele, a retinoblastoma results. The retinoblastoma gene is the best characterized tumour suppressor gene. Its product is a nuclear phosphoprotein which helps regulate DNA synthesis.

Associations 5% occur with a pineal or other tumour (=trilateral retinoblastoma). Secondary malignancies such as osteosarcoma and rhabdomyosarcoma are more frequent, and they are the main causes of death of patients with hereditary retinoblastoma. medline:10921315

Treatment There has been a trend away from enucleation (removal of the eye) + radiotherapy toward focal procedures to preserve eye and sight, if possible.

Enucleation may be needed with large tumours, long standing retinal detachments, and optic nerve invasion or extrascleral extension.

External beam radiotherapy has a rôle, but is associated with secondary non-ocular cancers in the field of radiation (eg in those carrying the RB-1 germline mutation).

Ophthalmic plaque brachytherapy has a more focal and shielded radiation field, and may carry less risk, but is limited to small to medium retinoblastomas in accessible locations.

Cryotherapy & transpupillary thermotherapy (TTT) can give control of selected small tumours. 'Chemoreduction' is achieved by iv or subconjunctival chemotherapy to allow TTT, cryotherapy & radiotherapy.

Haematopoietic stem cell rescue This may ↑ survival if distant metastasis—but CNS involvement means poorer prognosis. medline:10651386

Screening parents & siblings This is needed for accurate genetic counselling and to allow presymptomatic treatment. **Germ-line mosaicism** must be considered as a genetic transmission pattern. If a parent is germ-line mosaic, the possibility of bearing more babies with retinoblastoma is higher than conventionally believed. medline:10704567

Eye movements and squint

To maintain single vision, fine co-ordination of eye movement of both eyes is necessary. Abnormality of the co-ordinated movement is called squint. *Other names for squint:* strabismus; tropia. Exotropia is divergent (one eye turned out) squint; esotropia is (one eye turned in) convergent squint. Prominent epicanthic folds (diagram) may produce pseudosquint.

Non-paralytic squints These usually start in childhood. The range of eye movements is full. Squints may be constant or not. All squints need ophthalmological assessment as vision may be damaged if not treated.

Difficult, eg in uncooperative children. Screening tests:

- 1 Corneal reflection: reflection from a bright light falls centrally and symmetrically on each cornea if no squint, asymmetrically if squint present.
- 2 Cover test: movement of the uncovered eye to take up fixation as the other eye is covered demonstrates manifest squint; latent squint is revealed by movement of the covered eye as the cover is removed.

Convergent squint This is the commonest type in children. There may be no cause, or it may be due to hypermetropia (p 490). If the eye is left without focused input, vision may never develop satisfactorily in that eye (it has *amblyopia*).

Divergent squint These tend to occur in older children and are often intermittent. Amblyopia is less commonly a problem.

Management Remember 3 'O's: *Observe, Optic, Operate*. Treatment should start as soon as the squint is noticed. *Observe* Assess the refractive state of the eyes after cyclopentolate 1% eyedrops; the cycloplegia allows objective determination of the refractive state of the eyes; the mydriasis allows a good view into the eye to exclude abnormality, eg cataract, macular scarring, retinoblastoma, optic atrophy. Spectacles are then provided to correct refractive errors. *Optic* The good eye may be patched to encourage use of the one tending to squint. *Operate* (eg resection and recession of rectus muscles): These help alignment and give good cosmetic results. NB: use of botulinum toxin has helped some patients with esotropia (see p 522).

medline:9541431 & 11093096

Paralytic squint Diplopia is most marked when trying to look in the direction of pull of the paralysed muscle. When the separation between the two images is greatest the image from the paralysed eye is furthest from the midline and faintest.

Third nerve palsy Ptosis, proptosis (as recti tone ↓), fixed pupil dilatation, with the eye looking down and out. Causes: p 488.

Fourth nerve palsy There is diplopia and the patient may hold his head tilted (ocular torticollis). The eye looks upward, in adduction and cannot look down and in (superior oblique paralysed)* Causes: trauma 30%, diabetes 30%, tumour, idiopathic.

Sixth nerve palsy There is diplopia in the horizontal plane. The eye is medially deviated and cannot move *laterally* from midline, as the *lateral rectus* is paralysed. Tumour causing ↑intracranial pressure (compresses the nerve on the edge of the petrous temporal bone), trauma to base of skull, or vascular. Botulinum toxin can eliminate need for strabismus surgery in selected VI palsies.

medline.10489509

*Superior oblique and inferior oblique aid eye abduction (ie lateral rotation), while the superior and the inferior rectus adduct the eye. The superior oblique also lowers the gaze while the inferior oblique elevates it—S Ali *BMJ* 2002 i 962.

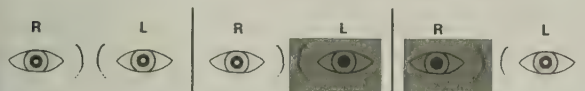
Best results in childhood strabismus are achieved by:

- Early detection of amblyopia. If >7yrs old, amblyopia may be permanent.
- Conscientious and disciplined amblyopia treatment.
- Optimal glasses (especially full plus in esotropia).
- Having the child see as straight as possible as soon as possible after amblyopia treatment is optimized. www.netspace.net.au/~strabism/strabimuslec.htm
- *Gobin's principles*: Evaluate *all* aspects of the strabismus (horizontal, vertical and oblique); search for the obstacles to ocular movements which cause alteration of binocular vision; remove them.

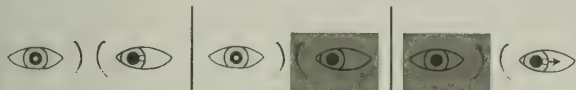
medline:1430799



PSEUDOSQUINT Wide epicanthic folds give the appearance of squint in the eye looking towards the nose. That the eyes are correctly aligned is confirmed by the corneal reflection.



NORMAL Corneal reflection shows correct alignment. Neither eye moves as they are alternately covered.



LEFT CONVERGENT SQUINT Corneal reflection shows malalignment. As the right eye is covered the left moves out to take up fixation.



LEFT DIVERGENT SQUINT Corneal reflection shows malalignment. As the right eye is covered the left eye moves in to take up fixation.

IB If there is **eccentric fixation** (ie foveal vision is so poor that it is not used for fixation), the deviating eye will not move to take up fixation. Corneal reflection shows that malalignment is present. The cover test relies on the ability to fixate.

Pupils

Pupil inequality Light detection by the retina is passed to the brain via the optic nerve (afferent pathway) and pupil constriction is mediated by the oculomotor (third) cranial nerve (efferent pathway). The sympathetic nervous system is responsible for pupil dilatation via the ciliary nerves.

Afferent defects (absent direct response) The pupil will not respond to light, but it will constrict to a beam in the other eye (consensual response). Constriction to accommodation still occurs. Causes: optic neuritis, optic atrophy, retinal disease. The pupils appear the same size (consensual response unaffected). The Marcus Gunn swinging flashlight test: shine a beam into the normal eye. Both pupils will constrict (direct and consensual reaction). Swing the light to the affected eye, and if the pupil dilates it is said to be a Marcus Gunn pupil.

Efferent defects The third nerve also mediates eye movement and eyelid retraction. With complete third nerve palsy there is complete ptosis, a fixed dilated pupil, and the eye looks down (superior oblique still acts) and out (lateral rectus acting). Causes: cavernous sinus lesions, superior orbital fissure syndrome, diabetes mellitus, posterior communicating artery aneurysm. The pupil is often spared in vascular causes, eg with diabetes and hypertension. Pupillary fibres run in the periphery, and are the first to be involved in compressive lesions by tumour or aneurysm.

Other causes of a fixed dilated pupil Mydriatics, trauma (blow to iris), acute glaucoma, coning ie uncal herniation (OHCM p 814).

Holmes-Adie pupil (typical patient) A young woman, with sudden blurring of near vision—as accommodation is partially paralysed. Signs: The pupil is slightly dilated, with a very slow response to accommodation, and, especially, to light (the light reflex may appear absent unless a prolonged, intense source is used). The Holmes-Adie syndrome entails this pupil pathology with absent knee and ankle jerks (selective impairment of monosynaptic connections of Ia afferents). medline:10431290

Horner's syndrome This occurs on disrupting the sympathetic nerve supply to the iris. The pupil is miotic (smaller) and there is partial ptosis of the lid, but normal range of lid movement (supply to levator palpebrae is by the third nerve). The pupil does not dilate in the dark. Loss of sweating usually indicates a lesion proximal to the carotid plexus—if distal, the sudomotor (*sudor* = sweat) fibres will have separated, so sweating is intact. Causes:

- Posterior inferior cerebellar artery or basilar artery occlusion
- Multiple sclerosis
- Hypothalamic lesions
- Syringomyelia in the pons
- Cavernous sinus thrombosis
- Aortic aneurysm
- Klumpke's paralysis (p 724)
- Cervical lymphadenopathy
- Pancoast's tumour

Argyll Robertson pupil This occurs in neurosyphilis and diabetes mellitus. There is bilateral miosis & pupil irregularity. There is no response to light but there is response to accommodation (the prostitute's pupil accommodates but does not react). The iris mesoderm is spongy, the pupils dilate poorly, and there may be ptosis.

Causes of light-near dissociation (–ve to light +ve to accommodation): Argyll Robertson pupil; Holmes-Adie syndrome; meningitis; alcoholism; tectal lesions, eg pinealoma; mesencephalic or thalamic lesions. The path from the optic tract to the Edinger-Westphal nucleus is disrupted but deeper cortical connections remain intact, so accommodation is spared.

Refraction

Refractive errors arise from disorders of the size and shape of the eye. Correct refraction depends upon the distance between the cornea and the retina, and the curvatures of the lens and cornea.

Myopia (near sightedness) The eyeball is too long. In any eye, the nearer an object comes to the eye, the further back the image falls. With myopic eyes a close object will focus on the retina (short sightedness). In order to focus the image from more distant objects (which focus too far forward), concave spectacle (or contact) lenses are needed. **Causes** Genetic (chromosome 18p & 12q) medline:9634508—also excessive close work in the early decades (not just at school) may lead to changes in the synthesis of mRNA and the concentration of matrix metalloproteinase, resulting in myopia. **NB:** when aboriginal peoples are exposed to western education rates of myopia rise from ~0 to Western levels (eg 50%). Animal models are characterizing the triggers of retinal and scleral growth—eg acetylcholine, dopamine, and glucagon. medline:10785617

In normal growth, changes in eyeball and lens curvature compensate for the eye getting longer as it grows, but in myopic children, cornea and lens do not compensate as growth occurs, and myopia worsens with age. Most do not become myopic until the age of about 6yrs (a few are born myopic). Myopia will then usually continue to worsen until the late teens, when changes stop below 6 dioptres in most people. So is important, therefore, for children with myopia to have their eyes regularly checked, as spectacle changes are to be expected, perhaps every 6 months. Avoid over-correction as this can make myopia worse. <http://bmj.com/cgi/content/full/324/7347/1195>

In later life, increasing myopia may indicate developing cataracts.

Rarely ($\leq 3\%$), myopia progresses above 6 dioptres (sometimes up to >20 dioptres). This has serious consequences later in life because secondary degeneration of the vitreous and retina can lead to retinal detachment, choroidoretinal atrophy and macular bleeding.

Spectacles, contact lenses, or LASIK, p 525. Bifocals prevent retinal blur if accommodative lag (blur promotes myopia). Don't just think: 'spectacles': when reading, ensure good lighting; advocate a balance of physical activity and reading. <http://bmj.com/cgi/content/full/324/7347/1195> & medline:11772601

astigmatism This is present if cornea or lens don't have the same degree of curvature in horizontal and vertical planes, so that the image of objects is distorted either longitudinally or vertically. Correcting lenses compensate accordingly. It can occur alone, or with myopia or hypermetropia.

hypermetropia This is due to too short an eye. Distant objects, when the eye is at rest (not accommodating) are focused behind the retina. The ciliary muscles contract, and the lens gets more convex to focus the object on the retina. This can produce tiredness of gaze, and sometimes, convergent squint in children. Hypermetropia is corrected by convex lenses to bring the image forward to focus on the retina.

presbyopia The eye is provided by nature with a convex lens for focusing near objects (when the lens is in its resting state, focus is on distant objects; ciliary muscle action reduces tension in the lens, allowing it get more convex—for close focusing). With age, the natural lens of the eye becomes stiffer and less easy to deform, so that focusing for close objects becomes less possible—hence the development of long sightedness with age (presbyopia) and the need for glasses for reading. These changes usually start in the lens at about 40 years and are complete by 60 years.

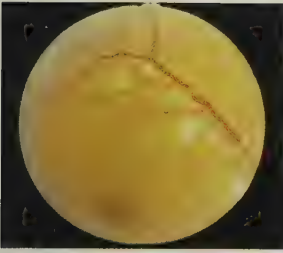


Plate 6.1 Cotton wool spots. Fluffy pale patches represent retinal microinfarcts, here following coronary artery bypass surgery.

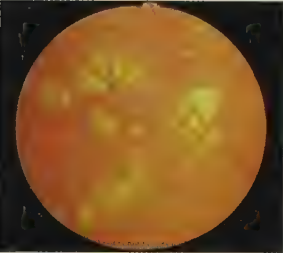


Plate 6.2 Hard exudates. Shiny pale hard exudates in a diabetic patient with type III hyperlipidaemia represent areas of lipid and protein leaked from plasma. The patient also had xanthomata.

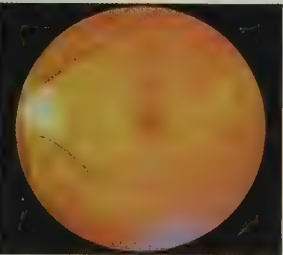


Plate 6.3 Retinal drusen. Pale lesions grouped round the fovea in an elderly patient are degenerative drusen which may precede disciform macular scarring.

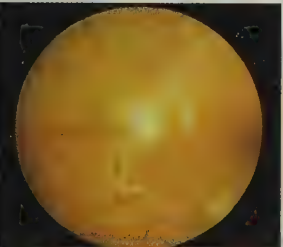


Plate 6.4 Retinal artery occlusion. Much of the retina is infarcted and hazy though the fovea is spared. The main retinal arterioles are sheathed. This patient had attacks of transient blindness before full occlusion.

Ophthalmic images

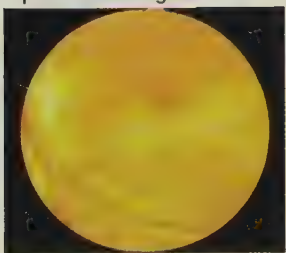


Plate 6.5 Retinal embolus. A small white embolus lodged in a branch retinal arteriole with surrounding area of pale retinal infarction. The embolus may be a fragment of atheromatous cholesterol or of cardiac valve.

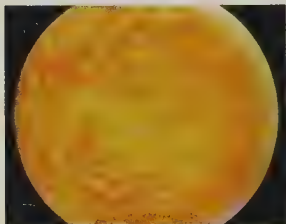


Plate 6.6 Retinal vein occlusion. Occlusion of the central retinal vein with a characteristic 'bloodstorm' appearance.

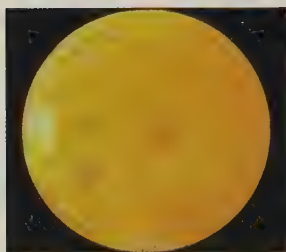


Plate 6.7 Diabetes, background retinopathy. There are scattered blot haemorrhages and sparse hard exudates but vision is normal.

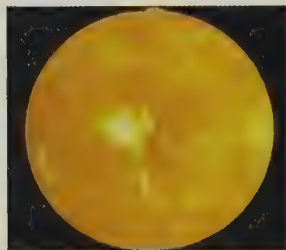


Plate 6.8 Diabetic maculopathy. Hard exudate at the fovea reduces vision irretrievably.

Images supplied by Peggy Frith, whom we here acknowledge and thank

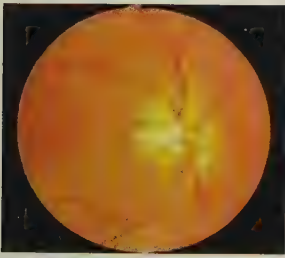


Plate 6.9 Diabetes, proliferative retinopathy. New vessels have formed in a looped fan on the inferior part of optic disc.

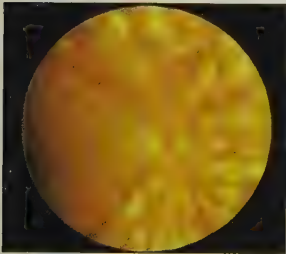


Plate 6.10 Treated proliferative retinopathy. Scars from laser burns outside the disc and major retinal vessel arcades appear after panretinal treatment. A tuft of disc new vessels persists at 7 o'clock.

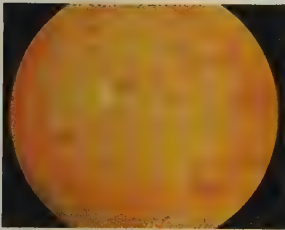


Plate 6.11 Accelerated hypertension. Flame haemorrhages, cotton wool spots, and disc swelling associated with renal artery stenosis. Mean blood pressure was 250/125 mmHg.

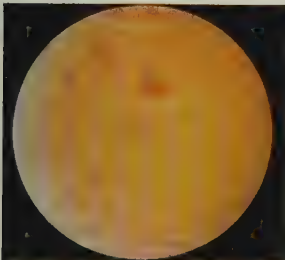


Plate 6.12 Retinal haemorrhages in leukaemia. Chronic granulocytic leukaemia with the white cell count of $76 \times 10^9/l$. The patient presented to his optician with visual blurring and the fundi showed widespread haemorrhages.

Ophthalmic images



Plate 6.13 Episcleritis. A sector of the eye shows dilated superficial vessels on the eye coat. The eye may sore and tender but is not painful.

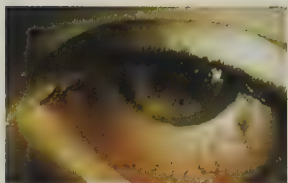


Plate 6.14 Scleritis. Scleritis with dilatation of both superficial and deep vessels and swelling of the eye coat. The eye is painful.



Plate 6.15 Optic disc in giant cell arteritis. The optic nerve head is infarcted and appears pale and swollen. In an elderly patient with a high ESR the diagnosis is giant cell arteritis.

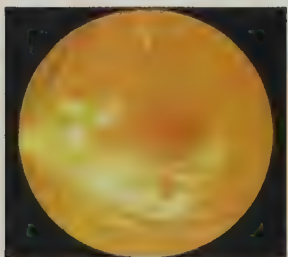
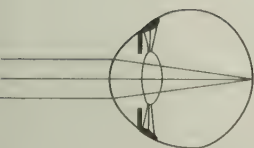


Plate 6.16 Retinitis in AIDS. The pale area with haemorrhages below the fovea is typical of cytomegalovirus retinitis in a patient with AIDS.

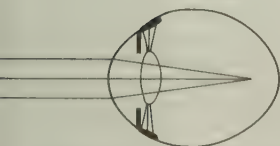


Normal eye



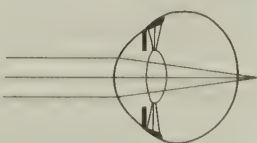
Normal eye

Myopia

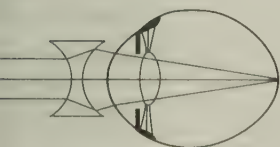


Light focused in front of retina

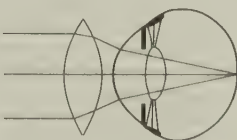
Hypermetropia



Light focused behind the retina



Corrected with concave lens



Corrected with convex lens

Visual field defects

When assessing for visual field defects establish 4 facts: 1 Is the defect bilateral? 2 Does the defect have sharp boundaries? 3 Do defects lie in the vertical or horizontal meridians? 4 What is the acuity? Lesions of sudden onset are often due to vascular causes. *Because of CNS plasticity, patients often think that the area of the defect (scotoma) is smaller than it is.* medline:9324542

Retinal lesions cause defects in one eye only and in the case of detachment and blood vessel occlusions (smaller vessels rather than the central ones) give defects with boundaries in the horizontal meridian. Field defects due to damage to the optic nerve tend to be central, asymmetrical, and unilateral—and acuity is often affected. When lesions are behind the optic chiasma field defects are bilateral, acuity tends not to be impaired (although objects in the affected field cannot be seen) and boundaries are in the vertical meridian.

Tests *Confrontation* The patient fixates on the examiner's eyes and notes the presence of a finger in all fields mapped, against the examiner's vision. It is used for testing peripheral fields. *Hat-pin confrontation* The patient fixates on a distant object, sitting about 1 metre away from the examiner. Red or white hat-pins are used to define any vertical meridian, the size of the blindspot and the boundaries of any scotomas. If a scotoma is 'absolute', the hat-pin disappears completely. If it is 'relative', the hat-pin only dulls as it moves across the field of the scotoma, as opposed to being bright and clear when in the unaffected field.

Optic chiasmal lesions may show a phenomenon whereby if 2 identical coloured objects are shown to one eye in the two vertical halves of the visual field (eg the right) one appears to be brighter and sharper than the other (if right eye, the left hemifield is brighter than the right). Computerized visual field analysers give accurate assessments of visual fields, with different programs delineating different lesions.

Amsler grid charts are used to look for distortion in central vision, which may be a sign of macular disease. The chart is 10 × 10cm square with 5mm squares drawn on it and a dot in the middle. With the chart held at 30cm the patient is instructed to look at the central dot and report any distorted squares or wavy lines (metamorphopsia).

Diagnosing the lesion's site Superior parts of the visual field fall inferiorly on the retina, temporal fields on the nasal retina and vice versa. Fibres from the nasal retina of both eyes cross in the optic chiasma to join uncrossed temporal retinal fibres. A pituitary tumour may disrupt the chiasma, affecting fibres crossing from nasal retinas, so causing bitemporal field defects. If it grows more to one side than the other, it can superimpose a central optic nerve defect as well. As fibres cross they maintain position (superior fibres stay superior). From the optic chiasma fibres pass in the optic tract to the lateral geniculate body, then as the optic radiation to the visual cortex.

A contralateral upper homonymous quadrantanopia may be caused by temporal lobe tumours. Posterior visual cortex lesions cause non-peripheral homonymous hemianopic scotomas (anterior visual cortex deals with peripheral vision)—with macular sparing, if the cause is posterior cerebral artery ischaemia (central areas have overlap flow via the middle cerebral artery which is why acuity may be preserved). MRI is the best aid to diagnosis.

• Ischaemia (TIA, migraine, stroke)
• Glioma • Meningioma • Abscess • AV malformation • Drugs, eg ciclosporin.

medline:8956125

Scotomata



Arcuate scotoma
—moderate glaucoma

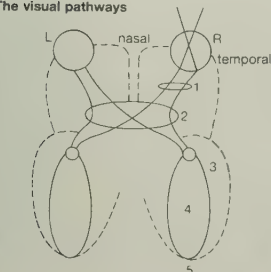


Unilateral defect
found with:
arterial occlusion
branch retinal vein
thrombosis
inferior retinal
detachment



Central scotoma
macular
degeneration or
macular oedema

The visual pathways



1 R optic nerve lesion
2 Lesion of chiasm—complicated
defects depending upon
which of the fibres are
most affected



3 Left superior quadrantanopia
due to R temporal lobe lesion



5 L homonymous scotoma
due to lesion at tip of R
occipital lobe



4 Homonymous hemianopia
from lesions affecting all R
optic radiation or visual cortex

Confrontation depends on mutual fixation, and it is necessary to concentrate hard to work out what is going on, but, for one second, allow yourself to reflect on what your patient will be thinking and feeling, most eloquently expressed by John Donne on accomplishing this manoeuvre:

*I fix mine eye on thine, and there
Pity my picture burning in thine eye. . .*

Towards the end of your professional life you will have engaged in mutual fixation many, many times, and as you continue to do so, ask yourself if these images of suffering are falling on your retina like so many layers of snow on a barren land, or whether, just sometimes, you might allow your eye to thaw and resolve itself into perhaps just one true tear.

The red eye

Red eyes are commonly also painful. Some causes are dangerous to vision and require specialist supervision (acute glaucoma, acute iritis, corneal ulcers); others are more easily treated (episcleritis, conjunctivitis, spontaneous conjunctival haemorrhage). Carefully examine all red eyes to assess acuity, cornea (use fluorescein drops p 496), and pupillary reflexes.

Acute closed angle glaucoma This is a disease of middle years or later life. The acute unocular attack is commonly preceded by blurred vision or haloes around lights—particularly at night-time. It is caused by blockage of drainage of aqueous from the anterior chamber via the canal of Schlemm. Dilatation of pupils at night exacerbates drainage block. Intraocular pressure then rises from the normal 15–20mmHg to 60 or 70mmHg.

Pain may be severe with nausea/vomiting; vision ↓; corneal oedema (haze); redness (circumcorneal); the pupil is fixed, dilated (may be vertically ovoid). ↑ Intraocular pressure may make the eye feel hard. A shallow anterior chamber (predisposing factor) may be seen in the other eye (shine a torch from the side, half the iris lies in shadow). If suspected send, to eye unit.

Pilocarpine 2–4% drops hourly (miosis opens the blocked, 'closed' drainage angle) + acetazolamide 500mg po stat (iv if vomiting) then 250mg/8h po. Acetazolamide ↓ formation of aqueous. IM morphine + prochlorperazine may be needed for pain. The patient must be admitted in the eye unit and the pressure must be monitored. Don't rely on newer agents alone, eg apraclonidine drops but they may have an additive effect when given with timolol and pilocarpine. medline:9380353 & 10366082

Peripheral iridectomy (laser or surgery) is done once intraocular pressure has been medically reduced (rarely as an emergency if medical management fails to control pressure). A small piece of iris is removed from the 12 o'clock position in both eyes to allow free circulation of aqueous.

Anterior uveitis The uvea is the pigmented part of the eye including iris, ciliary body, and choroid. The iris and ciliary body are called the anterior uvea and iris inflammation invariably involves the ciliary body too, so inflammation is best referred to as anterior uveitis. Remember to ask about systemic diseases (eg ankylosing spondylitis, sarcoid).*

Presentation is with pain of acute onset, photophobia, blurred vision (due to precipitates in the aqueous), lacrimation, circumcorneal redness (ciliary congestion), and a small pupil (initially from iris spasm; later it may be irregular or dilate irregularly due to adhesions). is +ve: pain increases as the eyes converge (and pupils constrict) as patients watch their finger approach their nose. medline:3676686 A slit lamp reveals white precipitates on the back of the cornea and cells in the anterior chamber. Sometimes sterile anterior chamber pus (hypopyon) may be seen.

It tends to afflict the young or middle-aged. There are many causes. Associations: joint problems (eg ankylosing spondylitis, Still's). It may relapse.

Refer to an ophthalmologist for treatment and follow-up. The aim is to prevent damage from prolonged inflammation (this can cause disruption of flow of aqueous inside the eye, with glaucoma occurring ± adhesions forming between iris and lens). Treatment is with steroids, eg prednisolone 0.5% drops every 2h, to reduce inflammation (hence pain, redness, and exudate). To prevent adhesions between lens and iris (synechiae) keep pupil dilated with cyclopentolate 0.5% 1–2 drops/6h, unless the iritis is very mild. Use the slit lamp to monitor inflammation.

*This is one of the few occasions when the Sherlock Holmes's among us can properly ask Have you been handling tarantulas recently? (their hairs cause uveitis).[@10832706]

Identifying the damaged eye

Ask yourself the following questions:

- 1 Is acuity affected? A quick but sensitive test is the ability to read newsprint with refractive errors corrected with glasses or a pin-hole. Reduced acuity suggests dangerous pathology.
- 2 Is the globe painful? Pain is potentially sinister, foreign body sensation may be so, irritation rarely is.
- 3 Does the pupil respond to light? Absent or sluggish response is sinister.
- 4 Is the cornea intact? Use fluorescein eyedrops, p 496. Corneal damage may be due to trauma or ulcers.

Enquire also about trauma and discharge, general health and drugs, and remember to check for raised pressure.

► If in doubt, obtain a specialist opinion today.

	<i>Conjunctivitis</i>	<i>Anterior uveitis</i>	<i>Acute glaucoma</i>
Pain	±	++	++ to +++
Photophobia	+	++	—
Acuity	normal	↓	↓
Cornea	normal	normal	steamy or hazy
Pupil	normal	small	large
Intraocular pressure	normal	normal	↑

More red eyes—cornea and conjunctiva

Corneal problems **Keratitis** is corneal inflammation (identified by a white spot on the cornea—indicating a collection of white cells in corneal tissue). **Corneal ulceration** is an epithelial breach; it may occur without keratitis, eg in trauma, when prophylactic antibiotic ointment (eg chloramphenicol) may be used. Ulceration with keratitis is called ulcerative keratitis and must be treated as an emergency—see below. There is pain, photophobia, and sometimes blurred vision. Non-infective corneal ulceration may result from a scratch from a contact lens, trauma, or previous corneal disease.

Diagnosis Use fluorescein drops & a bright light (ideally with a blue filter, shone tangentially across the globe) to aid diagnosis. Corneal lesions stain green (drops are orange and become more yellow on contact with the eye). Ulcers may be bacterial (beware pseudomonas: may progress rapidly), herpetic (simplex; zoster), fungal (candida; aspergillus), protozoal (acanthamoeba) or from vasculitis, eg in rheumatoid arthritis. Except for a simple abrasion, which can be treated with chloramphenicol eye ointment ± an eye pad, get expert help. Don't try treating ulcerative keratitis on your own: scarring and visual loss may occur. ▶ Refer the same day, as treatment depends upon the cause; delay may cause acuity↓. Anyone with corneal ulceration or stromal suppuration must have an urgent diagnostic smear/Gram stain and scrape by a specialist. Liaise with microbiologist about samples. **Treatment** p 497.

In early stages of ophthalmic shingles, use oral aciclovir (p 484). *H simplex* dendritic ulcers: p 480. Cycloplegics (p 518) may prevent iris adhesions.

Episcleritis (PLATE 6.13) Inflammation below the conjunctiva in the episclera is often seen with an inflammatory nodule. The sclera may look blue below engorged vessels, which can be moved over the area, unlike in scleritis, where the engorged vessels are deeper. The eye aches dully and is tender, especially over the inflamed area. Acuity is usually OK. Usually no cause is found, but it may complicate PAN/SLE and rheumatic fever. It responds to steroid drops (eg clobetasone butyrate 0.1%/6h). medline:10599674 &: 7028088

Scleritis (PLATE 6.14) Rarely, the sclera itself is inflamed. There is more generalized inflammation with oedema of the conjunctiva and thinning of the sclera (if very severe globe perforation is a risk). It may be associated with connective tissue disorders. Refer to a specialist. Most will need oral corticosteroids or immunosuppressives to control the disease. medline:11024419

Conjunctivitis ▶ Conjunctivitis is usually bilateral, if apparently unilateral consider other diagnoses (p 495), eg acute glaucoma. The conjunctiva is red and inflamed, and the hyperaemic vessels may be moved over the sclera, by gentle pressure on the globe. Acuity, pupillary responses, and corneal lustre are unaffected. Eyes itch, burn, and lacrimate. There may be photophobia. Purulent discharge may stick the eyelids together. The cause may be viral (highly infectious adenovirus)—small lymphoid aggregates appear as follicles on conjunctiva, bacterial (purulent discharge more prominent), or allergic. The affliction is usually self-limiting (although allergic responses may be more prolonged). In prolonged conjunctivitis, especially in young adults or those with venereal disease consider chlamydial infection—see ophthalmia neonatorum, p 100. **Treatment** Usually with eg chloramphenicol ointment at night. For chlamydia, get expert help.

For allergic conditions, try antihistamine drops, eg emedastine; refer if not settling in a few days. Sodium cromoglycate and steroid drops (after advice from an ophthalmologist) may help. medline:10337432

Subconjunctival haemorrhage This self-limiting but alarming-looking collection of blood behind the conjunctiva from a small bleed. Check BP.

Management of corneal ulcers

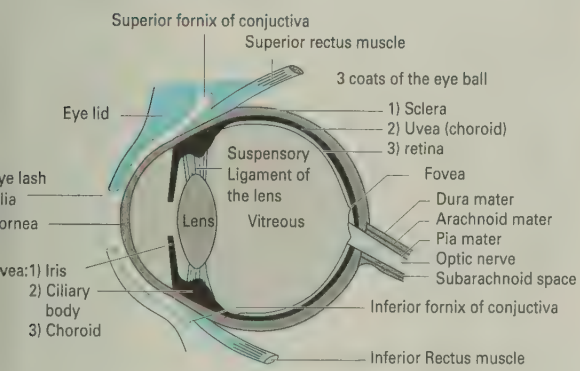
Refer to a specialist. What appears below is more so that one can have an intelligent conversation with a specialist, and should not be regarded as a blueprint for therapy by the inexperienced.

- Smears and cultures** Liaise with microbiologist. Specialists may take:
- 1 Smear for Gram stain—if chronic ulcer Giemsa, PAS (periodic acid Schiff) for fungi, ZN (Ziehl-Neelsen) or auromine for TB.
 - 2 Conjunctival swab to blood agar for tear film contaminants.
 - 3 Corneal scrape (by experienced person) from multiple areas of ulcer edge with needle for direct inoculation.
 - 4 Request the cultures detailed below.

Acute history: Presume bacterial, so culture with blood agar (grows most organisms), chocolate agar (for *Haemophilus* and *Neisseria*), nutrient broth (anaerobes), cooked meat broth (aerobes/anaerobes). **Chronic history.** Consider rarities; culture as above + BH (brain heart infusion broth for fastidious organisms and fungi), Sabouraud's plate for fungi, anaerobic blood agar (peptococcus, propionobacteria) + thioglycollate (anaerobes).

Unusual features: Use also viral transport medium, Lowenstein-Jensen agar slope for TB, *E Coli* seeded agar for acanthamoeba.

Management: Do smears & culture. Get expert help. Until these results are known, give either ciprofloxacin drops (or cefuroxime 0.3% drops with gentamicin drops). Adapt these in the light of culture results. Hospital admission may be needed, especially if there is diabetes or immunosuppression, or you doubt that the patient will manage his or her drops. Ciprofloxacin may be given up to 2 drops every 15min for 6h; then every 30mins for the next 18 hours; reducing to 2 drops hourly for day 2; then 2 drops 4-hourly for ≤ 12 days (maximum duration of therapy: 21 days).



Sudden painless loss of vision

This is an emergency, especially if there is: retinal artery occlusion of <6h; any sudden visual loss of <6h if the cause cannot be established—or giant cell arteritis (GCA). • **headache** associated? (GCA, eg aged >50yrs ± ESR↑); • **eye movements** painful? (optic neuritis); • **lights/flashes** preceding visual loss (retinal detachment); • **like a curtain** descending? (amaurosis fugax may precede permanent visual loss, eg from emboli or GCA; (PLATES 6.5 & 6.15). • **poorly controlled DM** (risks vitreous haemorrhage). Next, check acuity, pupil reactions, and fundi. Then refer.

Ischaemic optic neuropathy The optic nerve is damaged if the posterior ciliary arteries are occluded by inflammation or arteriosclerosis. **Fundoscopy:** pale, swollen optic disc.

It is vital to recognize this: the other eye is at risk until treatment is started. **Symptoms:** malaise, jaw claudication (chewing pain) ± tender scalp/temporal arteries (pulses may be absent). **Association:** polymyalgia. **Tests:** ESR >40 is suggestive; **temporal artery biopsy** may miss affected sections of artery. **Transcranial Doppler** may show microemboli. **Retinal changes:** PLATE 6.15. **Treatment:** start prednisolone 80mg/24h po promptly (some advocate higher iv doses if visual failure is occurring). Tailing off steroids as ESR and symptoms settle may take more than a year.

Associations: BP↑; lipids↑; DM. Treating these may protect vision in the other eye. Histology reveals necrosis and apoptosis at the photoreceptor level.

Central retinal artery occlusion (PLATE 6.4) There is dramatic visual loss within seconds of occlusion. In 90% acuity is finger counting or worse. An afferent pupil defect (p 488) appears within seconds and may precede retinal changes by 1h. The retina appears white, with a cherry red spot at the macula. Exclude temporal arteritis. Occlusion is often thromboembolic. Look for signs of atherosclerosis (bruits; BP↑), heart valve disease, diabetes, smoking, or lipids↑. **If seen within 1h of onset** apply firm pressure on the globe, increasing until the patient feels pain. Then suddenly release. This may dislodge an embolus, and propel it down a branch (PLATE 6.5). There is no reliable treatment, and if the occlusion lasts much longer than an hour the optic nerve will atrophy, causing blindness. If a single branch of the retinal artery is occluded, the retinal and visual changes relate only to the part of the retina supplied.

Vitreous haemorrhage This is a particularly common cause of visual loss in diabetics with new vessel formation. It may also occur in bleeding disorders, retinal detachment and with central retinal vein or branch vein occlusions. With a large enough bleed to obscure vision, there is no red reflex and the retina may not be seen. They undergo spontaneous absorption, and treatment is expectant for the haemorrhage itself, as well as being directed to the cause (eg photocoagulation of new vessels). If the vitreous haemorrhage is unresolved at 3 months, vitrectomy may be done to remove the blood in the vitreous. Small extravasations of blood produce vitreous floaters, (seen by the patient as small black dots or tiny ring-like forms with clear centres) which may not greatly obscure vision.

Unilateral ↓acuity occurs over hours or days. Discrimination of colour is affected: reds appear less red, 'red desaturation'—and eye movements may hurt. The pupil shows an afferent defect (p 488). The optic disc may be swollen (papillitis) unless the inflammation is central (retrobulbar neuritis). Recovery is usual over 2–6 weeks, but 45–80% develop multiple sclerosis (MS) over the next 15yrs. Other causes: neurosyphilis, other demyelinations (eg Devic's disease), Leber's optic atrophy, diabetes mellitus, and vitamin deficiency.

High-dose methylprednisolone for 3 days (250mg/6h iv), then oral prednisolone (1mg/kg/day) for 11 days appears to reduce the risk of developing MS in the next 2yrs, from ~15% to 7%. Seek expert advice.

Retinal vein occlusion: central or branch vein?

Central retinal vein occlusion (PLATE 6.6) Incidence increases with age. It is commoner than arterial occlusion. Causes: chronic simple glaucoma, arteriosclerosis, BP↑, and polycythaemia. If the whole central retinal vein is thrombosed, there is sudden visual loss (eg acuity reduced to finger counting). The fundus is like a 'stormy sunset' (the angry-looking red clouds are haemorrhages, beside engorged veins). There is also hyperaemia.

Long-term outcome is variable, with possible improvement for 6 months to one year; peripheral vision tends to improve most, leaving macular vision impaired. The main problems are macular oedema and neovascular glaucoma secondary to iris neovascularization. ~1/3 of eyes show significant non-perfusion on fluorescein angiography, of which one-half would develop neovascular glaucoma (called '100 days glaucoma' as it develops about this time after the occlusion). There is no validated treatment for post-occlusion macular oedema. medline:10654172 &: 8738462 Argon laser grid photocoagulation is used, but has yet to be fully validated. medline:1525880

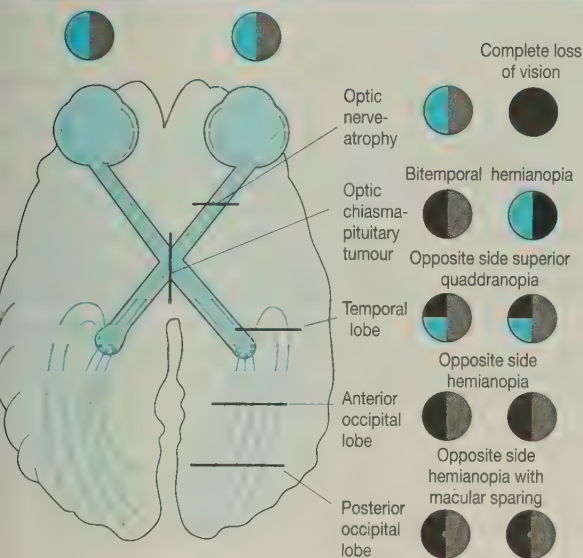
Branch retinal vein occlusion Unilateral visual loss and fundal appearances in the corresponding area. Retinal capillary non-perfusion can lead to retinal new vessel formation. Treatment of neovascularization (confirmed by fluorescein angiography) with laser photocoagulation, ↓risk of intraocular haemorrhage by 50%. Macular oedema persisting for months without improvement may receive grid pattern argon laser photocoagulation (± arterial crimping). medline:10990051

Diagnosis and differential diagnosis Doppler has a rôle in diagnosis. medline:11094189 Other causes of sudden loss of vision: ●Retinal detachment (p 506) ●Acute glaucoma (painful, p 494) ●Migraine

Stroke patients may complain of monocular blindness but visual field testing will usually reveal a homonymous hemianopia. Sudden bilateral visual loss is unusual (may be CMV infection in HIV patients, p 510).

Images relating to sudden loss of vision

www.bxt.co.uk/full.cfm?1089



Gradual loss of vision

Gradual loss of vision is mostly from cataract or age-related macular degeneration. It may also be due to choroiditis, a creeping inferior retinal detachment (p 506), or a choroidal melanoma. If the loss is bilateral (usually asymmetrical), think of chronic glaucoma (p 502), diabetic and hypertensive retinopathy (p 510), and optic atrophy (see below).

Be aware that for many, the chief question is likely to be 'will I go blind': be optimistic where possible. Patients may *not* tell you that they also fear they are going mad—having complex visual hallucinations, typically of faces. These occur without psychiatric morbidity and are often related to failing vision in the elderly: the Charles Bonnet syndrome. medline:9798740

Choroiditis (choroidoretinitis) The choroid is part of the uvea (iris, ciliary body & choroid), and inflammatory disorders affecting the uvea may also affect the choroid. The retina may be invaded by organisms which set up a granulomatous reaction (which can be mistaken for a retinoblastoma). Toxoplasmosis and toxocara are now more common than TB. Sarcoid is another cause.

Tests: CXR; Mantoux; serology. In the acute phase, vision may be blurred, a grey-white raised patch is seen on the retina, vitreous opacities occur, and there may be cells in the anterior chamber. Later, a choroidoretinal scar (white patch with pigmentation around) will be seen, these being symptomless unless involving the macula. Treat the cause.

Choroid melanomas are the commonest malignant tumour of the eye. Appearing as mottled grey/black on the fundus, they cause retinal detachment immediately over the growth. Spread is haematogenous or by local orbit invasion. *Treatment* Enucleation; local treatment is sometimes possible, eg sight- and globe-preserving stereotactic radiosurgery. medline:10974734

Age-related macular degeneration (ARMD) is the commonest cause of registrable blindness in the UK. It occurs in the elderly who complain of deterioration of central vision. There is loss of acuity; visual fields are unaffected. The disc appears normal but there is pigment, fine exudate, and bleeding at the macula. Drusen (PLATE 6.3) may precede these changes, and as early treatment (OPPOSITE) may save vision, observation of drusen is an important (but rather difficult) art to acquire. Rarely, the macula is oedematous and lifted by a big mass of exudate, called disciform degeneration. There is evidence that use of statins (OHCM p 708) **may** cause a 10-fold ↓ in risk of ARMD.*

Tobacco-alcohol amblyopia This is the result of toxic effects of cyanide radicals when heavy smoking and alcohol consumption are combined. Signs: optic atrophy; loss of red/green discrimination (an early sign); scotomata. Vitamins may help (B1, B2, B6, B12, folic acid). medline:8328548 & 4050326 & 8192611 & 3997034

Optic atrophy The optic disc is pale (but the degree of pallor doesn't correspond to visual loss). Optic atrophy may be secondary to ↑ intraocular pressure (in glaucoma), or retinal damage (as in choroiditis, retinitis pigmentosa, cerebromacular degeneration), or be due to ischaemia (as in retinal artery occlusion). Toxic causes other than tobacco are methanol, lead, arsenic, quinine, and carbon bisulfide. *Other causes* Leber's optic atrophy (p 752), multiple sclerosis, syphilis, external pressure on the nerve (intra-orbital or intracranial tumours, Paget's disease affecting the skull).

Use of services by older patients with failing vision

There is good evidence in the UK of under-use of services. Population-based cross-sectional studies in primary care show that prevalence of bilateral visual impairment (acuity $<6/12$) is ~30%. Most of these are not in touch with ophthalmic services. ▶ *Three-quarters of these have remediable problems.* In one study, 20% had acuity in one or both eyes of $<6/60$ ('blind'). Typical causes were found to be cataract, (30%), macular degeneration (8%), and undiagnosed chronic glaucoma.

medline~9603746

Managing age-related macular degeneration

Laser photocoagulation can halt progression, eg if choroidal neovascular lesions don't extend under the centre of the retina (photocoagulation destroys photoreceptors overlying abnormal vessels). Here *photodynamic laser therapy (OHCM p 720)* using the drug verteporfin may reduce vision loss.

medline:11094244

Screening: Once drusen are seen, screening needs to be one eye at a time, and very frequent, for signs of developing neovascularization: a simple method is to see if straight lines (on graph paper) appear straight? Sudden blank spots or distortions merit prompt referral.

medline~11110721

As getting neovascularization in one eye is associated with a 50% chance of developing it in the other, it is vital to save vision in either eye since one doesn't know which eye will end up being better. If these measures are inapplicable, most must rely on visual aids (eg to read). Zinc and β -carotene replacement are advocated by some. The best advice may be to take a diet rich in fruit and leafy green vegetables, with supplements if this diet causes problems.

medline:10796708

Chronic simple (open angle) glaucoma

Simple glaucoma is asymptomatic until visual fields are severely impaired; hence the need for screening—but most people found to have ↑ intraocular pressures (eg on 10% of routine NHS sight tests if >40yrs old) do not have glaucoma when their fundi and peripheral fields are charted (this can be a lengthy and tricky business). Once raised pressures are found, life-long follow-up is needed (at least yearly, and very much more often in the early stages)—which is why glaucoma accounts for such a heavy burden in ophthalmology clinics (eg 25% of the work), and why highstreet optometrists are being encouraged to take over at least part of this monitoring burden. Glaucoma accounts for 7% of new blind registrations.

Pathogenesis Intraocular pressure (IOP) ≥ 21 mmHg causes optic disc cupping \pm capillary closure, hence nerve damage, with sausage-shaped field defects (scotomata) near the blind spot, which may coalesce to form big defects. Nasal and superior fields are lost first (temporal last). Normal optic cups are similar in shape and occupy <50% of the optic disc. In glaucoma these enlarge (esp. along vertical axis). As damage progresses the optic disc pales (atrophy), and the cup widens and deepens, so the blood vessels emerging from the disc appear to have breaks as they disappear into the cup and are then seen at the base again (p 503). Since the central field is intact, good acuity is maintained, so presentation is often delayed until irreversible optic nerve damage. Control of IOP does stop visual field loss. medline:11024415 Some get glaucoma with normal IOP (eg if retrobulbar blood flow ↓). medline:11078851

High-risk Screen if high-risk: >35yrs old, +ve family history (esp. siblings); Afro-Caribbean; myopia; diabetic or thyroid eye disease. Technology is making primary prevention practical and cost-effective. Tests must be combined, as follows, in order of effectiveness: ● Multiple stimulus static visual field screening ● Optic disc cupping ● IOPs ↑. medline:10743748 & 10472959

Treatment The aim is to reduce intraocular pressure to <21 mmHg. Surgery is used if drugs fail. medline:10743748 & 10472959 Use twice daily (/24h for Timoptol LA[®]) to ↓ production of aqueous. They are β -blockers, so be cautious if asthma or heart failure. (Systemic absorption occurs with no first-pass metabolism by the liver.) SE: dry eyes, corneal anaesthesia, allergy, ↓ exercise tolerance. medline:7646670 Betaxolol may cause stinging (less if Betoptic-S[®] formulation is used—expensive). medline:9230824

507 **Prostaglandins** reduce resistance to the outflow of aqueous. It causes miosis and may cause blurring of vision and brow ache due to ciliary muscle spasm. Rarely, retinal detachment can occur. Presbyopes tolerate it better than the young and short sighted. Use 4 times daily. Because of these problems, pilocarpine is second-line treatment, or, with improving surgical options, some would say not even second line. (Pilogel[®]) reduces blurred vision as it is applied at night.

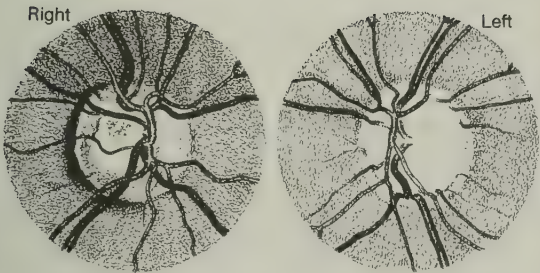
Adrenergics and **cholinergics** These sympathomimetics probably act by ↑ outflow via the trabecular meshwork and uveosclera. Caution if heart disease or if BP ↑; avoid in closed angle glaucoma. SE: sore, smarting, red eyes; blurred vision. Use once or twice daily.

Acetazolamide 250mg/24h–500mg/12h PO or topical (2%/8h) ↓ production of aqueous. (Cosopt[®]=dorzolamide with timolol; use twice daily). medline:10598693 SE of acetazolamide: lassitude, dyspepsia, K⁺ ↓, paraesthesia—so dorzolamide is preferred (but its SE are bitter taste, blurred vision, epiphora). Avoid both if pregnant.

Prostaglandins (Latanoprost) ↑ uveoscleral outflow. Dose: once daily. SE: brown iris pigmentation; uveitis. Said to be better than timolol. medline 11466259

Flap-valve trabeculectomy is a common operation. It probably acts as a drainage operation. Problems include early failure, and worsening of cataract (normal healing mitigates its effect, but this can be delayed by topical cytotoxics, such as fluoruracil). Effects of

is often short-term. It may have a rôle in older patients.



The two optic discs of a patient with open angle glaucoma which has not yet damaged the right optic disc. The left optic disc is grossly cupped and atrophic. (From J Parr *Introduction to Ophthalmology*, OUP.)

Cataract

► When a cataract is found, measure blood glucose (to exclude DM). ► Any opacity in the lens is called a cataract. ► The 4 major causes of blindness in the world are cataract, vitamin A deficiency, trachoma, and onchocerciasis. Cataracts are found in 75% of over 65s but in only 20% of 45–65-year-olds.

Ophthalmoscopic classification is by lens appearance. With immature cataracts the red reflex still occurs; if dense cataract there is no red reflex, or visible fundus. **Nuclear** cataracts change the lens refractive index and are common in old age, as are the cortical spoke-like wedge-shaped opacities. Anterior and posterior **posterior** cataracts are localized, are commonly inherited, and lie in the visual axis. **Subcapsular** opacities from steroid use are just deep to the lens capsule—in the visual axis. **Dispersed** opacities are common in normal lenses but are also seen in fast-developing cataracts in diabetes or dystrophia myotonica.

Presentation Blurred vision; unilateral cataracts are often unnoticed, but loss of stereopsis may affect distance judgment. Bilateral cataracts may cause gradual visual loss \pm frequent spectacle changes due to the refractive index of the lens changing, may cause dazzling—especially in sunlight, and may cause monocular diplopia. In children they may present as squint, loss of binocular function, as a white pupil, as nystagmus (infants), or as amblyopia.

Treatment The only treatment for cataract is surgery. Mydriatic drops, sunshades, and sunglasses may give some temporary help, but if symptoms are troubling, surgery is warranted. With modern surgery and lens implantation, visual acuity is not relevant. Consider surgery when symptoms restrict lifestyle, or if patients cannot see well enough to drive (the visual requirement is to read a car numberplate at 67 feet). Advise patients there is a slight risk to surgery: 2% have serious complications, and, although surgery usually greatly improves vision, the eye is not entirely normal afterwards. Many people continue to have a little dazzle of glare, and, for most, there is no effective near vision without spectacles. Distant spectacles are often required too. Remember one-third of people have co-existing ocular diseases, often macular degeneration, which may limit the outcome.

The ideal treatment is day case surgery using local anaesthesia with small incision surgery and phacoemulsion, combined with lens implantation. This is applicable to the vast majority of patients. Younger people, high myopes, and the squeamish may prefer a general anaesthetic. An incision of ~3mm is made, and the lens is removed by phacoemulsion (ultrasound breaks up the lens which is then aspirated into a cannula). The incision is then fractionally enlarged and an artificial lens (eg of perspex, acrylic, or silicon) implanted. Some lenses are foldable so they can be put through a smaller incision. The patient can usually return home immediately afterwards. A dressing may be needed for a few hours. With phacoemulsification, full activities can be resumed next day. With complicated surgery or extracapsular extraction, a larger incision is needed and there may be more limitations. Patients need to use antibiotic and anti-inflammatory eye drops for 3–6 weeks post-operatively. Then they need to change spectacles to get the full benefit of surgery.

Post-operative complications A very common event after surgery is thickening of the posterior capsule. This posterior membrane of the lens is deliberately left at surgery to make the surgery safer. In 5–30% of patients, over a few months to a few years post-operatively it opacifies. To the patient it seems as if the cataract is returning. It is easily treated by making a hole in the capsule (capsulotomy) with a Yag laser as an outpatient.

People often have a little eye irritation requiring additional or altered drops postoperatively. Some may have anterior uveitis requiring alternative medication. Rarely, there may be endophthalmitis, vitreous haemorrhage, retinal detachment, or glaucoma which may lead to permanent visual loss.

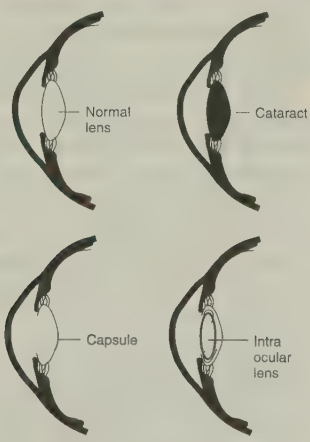
Cataract risk factors^{2,3}

Most cataracts are simply age related. In children, many are genetic (possibly some genetic influence in adults too). Cataracts may occur in diabetes mellitus; with topical, oral, or inhaled steroid use; with high myopia following trauma or other ocular surgery; and associated with systemic disorders such as dystrophia myotonica. In developing countries it may follow severe diarrhoeal illness and there may be an additional racial element.

Pre- and post-operative care

Prior to surgery ocular biometry should be undertaken. This is a measurement of the curvature of the cornea and the length of the eye which enables prediction of the suitable intraocular lens implant for the patient. In most cases it is aimed to leave the patient emmetropic (in focus for distance), or just slightly myopic, but this may vary considerably depending on patient preference and pre-existing refraction. It is not an exact science as the clinical measurements vary and many people do continue to wear spectacles postoperatively for a remaining refractive error. On obtaining new spectacles many will experience symptoms of imbalance; these should settle in 2–3 weeks. If they do not, recommend return to the optometrist to check the correct spectacles have been dispensed. If there is still a continuing problem they should return to the ophthalmologist.

If patients develop a painful red eye or loss of vision post-operatively they should be referred back to the ophthalmologist urgently to deal with possible complications. Many experience awareness of the eye or dry, or gritty sensations and lubricants such as Viscotears® may help. If vision deteriorates with time they should initially visit the optometrist to see if they need a new prescription change; and thereafter the ophthalmologist to consider Yag laser capsulotomy or exclusion of other problems.



We thank Mr R Williams FRCS for preparing this page
 1 K Sundelin 1999 *J Cat Refract Surg* 25 246 2 G Johnson 1998 *Lancet* 351 925
 3 R Cumming 1998 *E-BM* 3 24 (if lifetime dose of inhaled beclometasone >2000mg)
 4 J Javitt 2000 *Ophthalmology* 107 2040 [@11054329]

The retina

Anatomy The retina consists of an outer pigmented layer (in contact with the choroid), and an inner sensory layer (in contact with the vitreous). At the centre of the posterior part lies the macula (the centre of which is termed the fovea), appearing yellowish and slightly oval. This has many cones, so acuity is greatest here. ~3mm medial to the fovea is the optic disc, which contains no rods or cones (the visual field's blind spot).

Optic disc Think colour; contour; cup. Colour should be a pale pink. It is more pallid in optic atrophy (p 500). Contour: the disc may appear oval in astigmatic eyes, and appear abnormally large in myopic eyes. Disc margins are blurred in papilloedema (eg from raised intracranial pressure, malignant hypertension, cavernous sinus thrombosis), and with optic neuritis. Blood vessels radiate away from the disc. The normal arterial/venous width ratio is 2:3. Venous engorgement appears in retinal vein thrombosis (PLATE 6.6); abnormal retinal pallor with artery occlusion (PLATE 6.4); and haemorrhages with exudates in hypertension and DM (PLATE 6.8 & 6.11). Cup: the disc has a physiological cup which lies centrally and should occupy ~1/3 of the diameter of the disc. Cup widening and deepening occurs in glaucoma (p 502).

Retinal detachment This may be 'simple', idiopathic, secondary to some intraocular problem (eg melanoma, or fibrous bands in the vitreous in diabetes), occur after cataract surgery, or result from trauma. Myopic eyes are more prone to detachment, the higher the myopia, the greater the risk.

In simple detachment, holes in the retina allow fluid to separate the sensory retina from the retinal pigment epithelium.

Detachment presents with 4 'f's: floaters, flashes (eg in 50%), field loss, and fall in acuity—painless and may be as a curtain falling over the vision (the curtain falls down as the lower half of the retina detaches upwards). Field defects indicate position and extent of the detachment (in superior detachments field loss is inferior). Ophthalmoscopy: grey opalescent retina, ballooning forward. Detachment of the lower half of the retina tends not to pull off the macula, but upper half detachments do. If it does detach, central vision is lost and doesn't recover completely even if the retina is successfully fixed. Rate of detachment varies: upper halves are quicker.

Urgent referral, eg for scleral silicone implants, cryotherapy, pneumatic retinopexy, or argon or laser coagulation to secure the retina. Post-op redetachment may occur (in 5–10%).

Retinitis pigmentosa This is the most prevalent inherited degeneration in the retina. Sporadic types are the most common, many among these being autosomal recessive, and there will be some newly-appearing autosomal dominant forms (AD). AD patients have the best prognosis; X-linked is the rarest form, with the worse prognosis. Through life, 25% retain the ability to read, albeit with much reduced visual fields. Only a few have acuity $\leq 6/60$ by the time they are 20; but by the age of 50, many are reduced to this level.

medline:11151217

Retinoblastoma See p 485.

Rothmann This obligate intracellular protozoan sometimes causes bilateral typical 'punched-out' heavily pigmented chorioretinal scars which can be seen on routine fundal examination. If scars involve the macular area there will be dramatic loss of acuity. OHCM p 576.

Macular holes

Synonyms Macular cyst, retinal hole, retinal tear, retinal perforation.

A macular hole is a full-thickness defect of retinal tissue (from the internal limiting membrane to the outer segment of the photoreceptor layer). It involves the fovea (visual acuity↓). Prevalence: eg 3.3/1000 in persons >55yrs. There is a 12% chance of a similar hole developing in the other eye.

Presentation Distorted vision with visual loss. Look for a tiny punched out area in the centre of the macula: there may be yellow-white deposits at the base. Slit lamp exam (with an examination contact lens) shows a round excavation with well-defined borders interrupting the beam of the slit lamp. Often there is an overlying semitranslucent tissue suspended over the hole—typically surrounded by a grey halo of detached retina.

➤ Refer for urgent ophthalmological assessment in eye casualty.

Cause Usually idiopathic (<10% have a history of trauma). There may be age-related focal shrinkage of the prefoveal vitreous cortex, causing tangential traction on the fovea (hence detachment and a macular hole).

Stage 1a Yellow spot (presumably intraretinal xanthophyll pigment, which has become more visible due to the foveolar detachment). This is not specific for macular hole: it can be associated with central serous chorioretinopathy, cystoid macular oedema, and solar maculopathy.

Stage 1b Occult hole: donut-shaped yellow ring (~200–300µm) centred on the foveola. ~50% of holes progress to stage 2.

Stage 2 Full thickness macular hole (<400µm). Prefoveal cortex usually separates eccentrically creating a semitransparent opacity, often larger than the hole, and the yellow ring disappears. These generally progress to stage 3.

Stage 3 Holes >400µm associated with partial vitreomacular separation.

Stage 4 Complete vitreous separation from the entire macula and disc.

Tests Fluorescein angiography (FA) useful for differentiating macular holes from cystoid macular oedema and choroidal neovascularization (CNV). This typically shows a window defect early in the angiogram that does not expand with time, and there is no leakage or accumulation of dye. There may be Amsler grid abnormalities (plotting small central scotomas is hard).

Treatment In rare cases the hole may close itself. Vitrectomy may relieve traction on the edge of the hole. The anterior and middle vitreous and internal limiting membrane are removed, an air bubble is introduced to nudge (tamponade) the macula back into position, which may work provided the patient spends the 1–2 post-op weeks face down (a great part of success is dependent on this). Vitrectomy with fluid–gas exchange for stage 2, 3, and 4 holes improves vision (compared with conservative treatment). Series of patients have been variously reported, with hole closure rates of ≥95% (~67% improve by 2 or more lines on visual acuity). Success is also possible if the hole is long-standing (>6 months) or if aged >80.

However, in some cases more than one operation is required to close the hole, and adverse effects may occur: macular retinal pigment epithelium changes, retinal detachments, iatrogenic retinal tears, enlargement of the hole, macular light toxicity, postoperative intraocular pressure spikes. Many patients develop cataracts (76% of cases requiring extraction within 2 years).

Patients not suitable or not wishing for this treatment need visual aids (eg to read).

The eye in diabetes mellitus

Diabetes can be bloody and blinding—the leading cause of blindness in those aged 20–65 (uk). Almost any part of the eye can be affected: cataract and retinopathy are the chief pathologies. 30% of adults have ocular problems when diabetes presents. At presentation, the lens may have a higher refractive index (possibly due to dehydration) producing relative myopia. On treatment, the refractive index reduces, and vision is more hypermetropic—so do not correct refractive errors until diabetes is controlled.

Structural changes Diabetes accelerates formation and progress of age-related cataract. Most commonly this is a premature senile cataract, but young diabetics can also be affected at presentation. In this case, the lens has taken up a lot of glucose which is converted by the enzyme aldolase reductase to sorbitol. Sorbitol cannot diffuse through the lens but increases the osmotic pressure within the lens; water is then taken up which can cause the lens fibres to rupture and a cataract forms thereafter. Rarely, the iris may be affected by diabetes, with new blood vessel formation on it (rubeosis), and, if these block the drainage of aqueous fluid, glaucoma can be caused.

Retinopathy Pathogenesis Microangiopathy in capillaries, precapillary arterioles and venules causes occlusion ± leakage. **Vessel occlusion** Causes ischaemia which leads to new vessels in the retina, optic disc, and iris, ie **proliferative retinopathy** (PLATE 6.9). New vessels can bleed, causing vitreous haemorrhage. As new vessels carry along with them fibrous tissue, retraction of this tissue increases risk of retinal detachment. Occlusion also causes **cotton wool spots** (PLATE 6.1, ischaemic nerve fibres). **Vascular leakage** As pericytes are lost, capillaries bulge (microaneurysms) and there is oedema & hard exudates (PLATE 6.2 & 6.8, made up of lipoprotein & lipid filled macrophages). Rupture of microaneurysms, at the nerve fibre level causes flame shaped haemorrhages (PLATE 6.11); when deep in the retina, **retrolental haemages** (PLATE 6.7) form.

► Pre-symptomatic screening enables laser photocoagulation (PLATE 6.10). Screen by regular eye exam or retinal photography. In rural areas, mobile screening has a rôle. medline8443485 Lesions are mostly at the posterior pole and can be easily seen by ophthalmoscope. **Background retinopathy** (PLATE 6.7) comprises microaneurysms (seen as 'dots'), haemorrhages (flame shaped or 'blots') and hard exudates (yellow patches, PLATE 6.2 & 6.8). Vision is normal. Background retinopathy can progress to sight-threatening maculopathy ± proliferative retinopathy. **Maculopathy** Leakage from the vessels close to the macula cause oedema & maculopathy. **Proliferative retinopathy**: Fine new vessels appear on the retina. Engorged tortuous veins, cotton wool spots (PLATE 6.1), large blot haemorrhages & vitreous haemorrhage.

► Refer those with maculopathy or proliferative retinopathy urgently to an ophthalmologist for treatment (eg photocoagulation) to protect vision.

Treatment Good control of diabetes can prevent new vessel formation. Concurrent diseases may accelerate retinopathy (eg hypertension, renal disease, pregnancy, and anaemia). Treat these (as appropriate), and hyperlipidaemia. Photocoagulation by laser is used to treat both maculopathy and proliferative retinopathy. Definite indications for photocoagulation are new vessels on the optic disc and vitreous haemorrhage. If vitreous haemorrhage is massive and does not clear, vitrectomy may be needed.

CNS effects Ocular palsies may occur, typically nerves III and VI. In diabetic third nerve palsy the pupil may be spared as fibres to the pupil run peripherally in the nerve, receiving blood supply from the pial vessels. Argyll Robertson pupils and Horner's syndrome may also occur (p 488).

The eye in systemic disease

Systemic disease often manifests itself in the eye and, in some cases, eye examination will first suggest the diagnosis.

Macular retinopathy This may be *arteriopathic* (arteriovenous): arteries nip veins where they cross—they share the same connective tissue sheath) or *hypertensive* (PLATE 6.11)—arteriolar vasoconstriction and leakage—producing (PLATE 6.2 & 6.8), and, rarely, . Thick, shiny arterial walls appear like wiring (called 'silver' or 'copper'). Narrowing of arterioles leads to localized infarction of the superficial retina seen as cotton wool spots (PLATE 6.1) and flame haemorrhages (PLATE 6.11). Leaks from these appear as hard exudates ± macular oedema/papilloedema (rare).

Emboli to the retina cause *amaurosis fugax* ('a curtain passing across the eyes', PLATE 6.5). *Typical cause*: atheroma (listen to carotids)*. Arrange urgent Doppler; treat eg by aspirin ± carotid endarterectomy.

Retinal haemorrhages are seen in leukaemia (PLATE 6.12); comma-shaped conjunctival haemorrhages and retinal new vessel formation may occur in sickle-cell disease; optic atrophy in pernicious anaemia.

Note also Roth spots (retinal infarcts) of infective endocarditis (OHCM p 136).

Metabolic disease Diabetes: p 508. Hyperthyroidism, and exophthalmos: OHCM p 295. In myxoedema, eyelid and periorbital oedema is quite common. Lens opacities may occur in hypoparathyroidism. Conjunctival and corneal calcification may occur in hyperparathyroidism. In gout, monosodium urate deposited in the conjunctiva may give sore eyes.

Granulomatous disorders (TB, sarcoid, leprosy, brucellosis, and toxoplasmosis) can all produce inflammation in the eye (uveitis). TB, congenital syphilis, sarcoid, CMV, and toxoplasmosis may all produce choroidoretinitis. In sarcoid there may be cranial nerve palsies.

Collagen diseases These also cause inflammation. Conjunctivitis is found in SLE and Reiter's syndrome; episcleritis in polyarteritis nodosa and SLE; scleritis in rheumatoid arthritis; and uveitis in ankylosing spondylitis and Reiter's syndrome (OHCM p 406). In dermatomyositis there is orbital oedema with retinal haemorrhages.

Keratoconjunctivitis sicca Sjögren's syndrome, (OHCM p 734) There is reduced tear formation (Schirmer filter paper test), producing a gritty feeling in the eyes. Decreased salivation also gives a dry mouth (xerostomia). It occurs in association with collagen diseases. Treatment is with artificial tears ('tears naturale', or hypromellose drops).

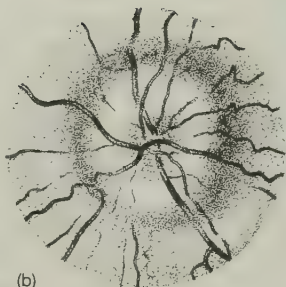
AIDS Those who are HIV+ve may get CMV retinitis (PLATE 6.16), with retinal spots ('pizza pie' fundus, signifying superficial retinal infarction) + flame haemorrhages involving more and more of the retina. This may be asymptomatic or cause blindness; it implies AIDS & ↓CD4 count. IV ganciclovir or its prodrug (oral valganciclovir) are used. medline.11948271
Cotton wool spots (PLATE 6.1) may indicate HIV retinopathy (may present before the full picture of AIDS); it is a microvasculopathy, not a retinitis. Candidiasis of the aqueous and vitreous is hard to treat. Kaposi's sarcoma may affect the lids or conjunctiva.

*Causes of amaurosis fugax: giant cell arteritis, orbital schwannomas, meningiomas, ocular small vessel disease, Churg–Strauss vasculitis, antiphospholipid syndrome, arrhythmias, dysfibrinogenemia, uveitis–glaucoma–hyphaema syndrome post-cataract extraction.

medline.11039076 & 11151281 & 10424154 & 10321544 & 10220729 & 9924923 & 9711117 & 9667576



(a)



(b)

(a) Papilloedema from raised intracranial pressure. The disc is swollen forwards and also outwards into the surrounding retina. The disc margin is completely hidden and in places retinal vessels are concealed, because edema has impaired the translucency of the disc tissues. The retinal veins are congested and there are a few haemorrhages. (b) Early papilloedema. The vessels crossing the disc are obscured in places due to the loss of translucency of the prelaminar tissue in which they lie. (From J Parr *Introduction to Ophthalmology*, oup.)

Tropical eye disease

Trachoma Caused by *Chlamydia trachomatis* (serotypes A, B, and C), this disease is spread mainly by flies—typically where it is hot, dry, and dusty and the people are poor, living near to their cattle. 400 million people are affected (100,000,000 are children).

Stage 1 There is lacrimation. Follicles under the upper lid give a fine granular appearance. **2** There is intense erythema. The follicles are larger and underneath both lids. A fine pannus and capillaries grow down towards the cornea. **3** The follicles rupture and are replaced by scar tissue. The pannus is more advanced. The cornea may ulcerate. **4** Scar tissue distorts the lids and causes entropion. Eyelashes scratch the cornea, which ulcerates.

Tx: Mass antitrachoma treatment: tetracycline 1% eye ointment 12-hrly for 5 days each month for 6 months. In active disease use 8-hrly for 6 weeks + sulfadimethoxine 1g stat, then 500mg/24h po for 10 days (erythromycin if a child). Single-dose azithromycin (20mg/kg) is effective. medline:10772587

Prophylaxis: Eg lid margin splitting + tarsal plate fracture + everting sutures.

Prevention: Good water/sanitation. Regular washing of faces. medline:10995092

Onchocerciasis (river blindness) This is caused by the microfilariae of the nematode *Onchocerca volvulus*, transmitted by black flies of the Simulium species. Of the 20–50 million people affected, 95% live in Africa. In some areas it may cause blindness in 40% of the population. Unless the eye is affected, problems are mostly confined to the skin. Fly bites result in nodules from which microfilariae are released. These eventually invade the eye, mainly the conjunctiva, cornea, ciliary body, and iris, but occasionally the retina or optic nerve. Sometimes they may be seen swimming in the aqueous or lying dead in the anterior chamber. The microfilariae initially excite inflammation; fibrosis then occurs around them. Reaction around the dead microfilariae in the cornea causes corneal opacities (nummular keratitis). Chronic iritis causes synechiae formation and may precipitate cataracts. The iris may become totally fixed. **Tx:** Skin snip tests, triple-antigen serology, polymerase chain reaction. **Prophylaxis:** Get expert help. Ivermectin is currently the chief microfilaricide, ~150µg/kg po as a single dose every 6–12 months, until adult worms die (OHCM p 617). In lightly infected expatriates, the first 3 doses are recommended to be monthly, with observation in hospital after the first dose (reactions are common in expatriates). Macrofilaricidal drugs are currently being developed for human use. medline~11734070

Xerophthalmia and keratomalacia These are manifestations of vitamin A deficiency, eg if weaned early on to vitamin A deficient milk products, or toddlers who eat few vegetables. Peak incidence: 2–5yrs; 40 million children worldwide. Night blindness and dry conjunctivae (xerosis) occur early. The cornea is unwettable and loses transparency. Small grey plaques (Bitot's spots) are commonly found raised from interpalpebral conjunctiva. Vitamin A reverses these changes. Early corneal xerosis is reversible. Corneal ulceration and perforation can occur. In keratomalacia there is massive softening of the cornea ± perforation and extrusion of the intraocular contents. **Tx:** For children: retinol palmitate 50,000u IM monthly until the eyes are normal (adult dose 100,000u, weekly to start with); or oral retinyl palmitate 200,000u po. β-carotene (a provitamin) 1.2×10^6 u po, is as effective, and cheaper. Avoid vitamin A in pregnancy (vitamin A embryopathy).

Eye trauma

Prevention is the key, eg wearing goggles, or plastic glasses when near small moving objects or using tools (avoids splinters, fish-hooks, and squash-ball injuries). *Always record acuity* (both eyes; if the uninjured one is blind take all injuries very seriously). Take a detailed history of the event.

If unable to open the injured eye, instil a few drops of local anaesthetic (tetracaine 1%): after a few mins, comfortable opening may be possible. Examine lids, conjunctiva, cornea, sclera, anterior chamber, pupil, iris, lens, vitreous, fundus, and eye movement. An irregular pupil may mean globe rupture. Afferent pupil defects (p 488) do not augur well for sight recovery. Note pain, discharge, or squint. CT may be very useful (foreign bodies may be magnetic, so avoid MRI).

Penetrating trauma Refer urgently: delays risk of ocular extrusion or infection. With uveal injury there is risk of sympathetic ophthalmia in the other eye. A history of flying objects (eg work with lathes, hammers, and chisels) should prompt careful examination and x-ray to exclude intraocular foreign bodies. Don't try to remove a large foreign body (knife; dart). Support the object with padding. Transport supine. Pad the *unaffected* eye to prevent damage from conjugate movement. Consider skull x-ray or CT to exclude intracranial involvement.

Foreign bodies in eye Have a low threshold for getting help; FBs often hide, so examine *all* the eye; for lid eversion see www.medinfo.ufl.edu/year1/bcs/clist/index.htm. FBs cause chemosis, subconjunctival haemorrhage, irregular pupils, iris prolapse, hyphaema, vitreous haemorrhage, and retinal tears. If you suspect a metal FB, x-ray the orbit. With high-velocity FBs, consider orbital ultrasound: pick-up rate is 90% vs 40% for x-rays—but skill is needed (not always available in busy A&E departments). Removal of superficial foreign bodies may be possible using a triangle of clean card. Use gentamicin 0.3% drops afterwards to prevent infection.

Corneal abrasions (Often from small fast-moving objects, eg children's finger-nails; twigs.) They may cause intense pain. Apply a drop of local anaesthetic, eg 1% tetracaine before examination. They stain with fluorescein and should heal within 48h. Apply gentamicin eye ointment, and pad the eye. Send the patient home with analgesics. Re-examine after 24h. If still having a foreign body sensation after removing the pad, stain again with fluorescein. If the cornea stains, repeat the procedure for another 24h. If it still staining after 48h, refer. NB: meta-analyses of small corneal abrasions do not favour using pads. medline:9789511 & 8224310 & 1119012

Burns Treat chemical burns promptly: instill anaesthetic drops (tetracaine) every 2min till the patient is comfortable. Then hold lids open and bathe the eyes in copious clean water while the specific antidote is sought. Often the patient will not hold the eye open due to excruciating pain. All burns may have late serious sequelae, eg corneal scarring, opacification, and lid damage. Alkali burns are more serious than acid.

Flash burns Welders and sunbed users who don't wear protection against uv light may damage corneal epithelium. There is a foreign body sensation, watering, and blepharospasm. Instill local anaesthetic drops every 2min. After the second application, excruciating pain can disappear like magic. Apply an antibiotic ointment, pad the eye, and it will recover in 24h. It is a very painful condition so be generous with analgesia.

Finally, remember to think of **trauma** in trauma patients who suddenly complain of visual problems.

Contusions and intraocular haemorrhage

Our eyes are protected by our bony orbital ridges. Severe contusions from large objects may damage the eye, but smaller objects such as champagne corks, squash balls, and airgun pellets (p 679) cause local contusion, eg resulting in lid bruises and subconjunctival haemorrhage (if the posterior limit of such a haemorrhage cannot be seen, consider fracture of the orbit). Both usually settle in 2 weeks. ▶Any injury penetrating the eyeball should receive immediate specialist treatment.

Anterior haemorrhage This usually affects acuity and should receive specialist attention. Blood is often found in the anterior chamber (hyphaema): small amounts clear spontaneously but if filling the anterior chamber, evacuation may be needed. It is often recognizable by pen-torch examination. Even small hyphaemas must be carefully evaluated (∴ refer): it may signify serious injury. *Late complications:* Glaucoma; corneal staining. Pain suggests glaucoma or re-bleeding.

Secondary haemorrhage This may occur within 5 days and may produce sight-threatening secondary glaucoma. Sometimes the iris is paralysed and dilated due to injury (called traumatic mydriasis). This usually recovers in a few days but sometimes it is permanent. Vitreous haemorrhage will cause dramatic fall in acuity. There will be no red reflex on ophthalmoscopy. Lens dislocation, tearing of the iris root, splitting of the choroid, detachment of the retina, and damage to the optic nerve may be other sequelae; they are more common if contusion is caused by smaller objects rather than large.

Blows to the orbit Blunt injury (eg from a football) can cause a sudden increase in the pressure within the orbit, and may cause blowout fractures with the orbital contents herniating into the maxillary sinus. Wethering of the inferior rectus and inferior oblique muscles causes diplopia. Test the sensation over the skin of the lower lid. Loss of sensation indicates infraorbital nerve injury, confirming a blowout fracture. CT may show the depressed fracture of the posterior orbital floor. Fracture reduction and muscle release is necessary. medline:8628546

Blindness and partial sight

The pattern of blindness around the world differs considerably, depending on local nutrition and economic factors. 90% of the world's blind live in developing countries—and 80% would not be blind if trained eye personnel, medicines, ophthalmic equipment, and patient referral systems were optimized.¹ The diseases responsible for most of the blindness in the world are trachoma, cataract (50% of the world's blindness), glaucoma, keratomalacia, onchocerciasis, and diabetic retinopathy. In the past smallpox, gonorrhoea, syphilis, and leprosy (10% of those affected were blind) were also common causes of blindness, but are less so now.

Rates of blindness are higher than 10/1000 in some parts of Africa and Asia, but in the UK and the USA rates are 2/1000. Blindness may be voluntarily registered in England, registration making one available for certain concessions. Although the word blind suggests inability to perceive light, a person is eligible for registration if their acuity is less than 3/60, or >3/60 but with substantial visual field loss (as in glaucoma). There are ~350,000 people registered blind in the UK. *Criteria for partial sight registration: acuity is <6/60 (or >6/60 with visual field restrictions).*

Causes of blindness These have changed considerably in the UK over the last 70 years. Whereas in the 1920s ophthalmia neonatorum (p 10) was responsible for 30% of blindness in English blind schools, this is now a rare but treatable disease. Retrolental fibroplasia was common in the 1950s, mostly affecting premature infants: monitoring of intra-arterial oxygen in premature babies tries to prevent this. With an ever-aging population, the diseases particularly afflicting this population are the common causes of blindness. Nearly two-thirds of the blind population are over 65 years of age, and nearly half over 75. Macular degeneration, cataract, and glaucoma are the three commonest causes of blindness.

Registration In England, responsibility for blind registration lies with the local authority. Application is made by a consultant ophthalmologist and is voluntary, not statutory. Registration as blind entitles one to extra tax allowances, reduced TV licence fees, some travel concessions, and access to talking books. Special certification from an ophthalmologist is necessary for the partially sighted to receive talking books. At one time it was statutory that the registered blind should receive a visit from a social worker but this is no longer the case, although the social services employ social workers who specialize in care of the blind. The Royal National Institute for the Blind² will advise on aids, such as guide dogs (available if required for employment).

Special educational facilities These provide for visually handicapped children. Special schools have a higher staff/pupil ratio, specialized equipment, and many have a visiting ophthalmologist. The disadvantage is that the children may not mix much with other children—especially if they are on the board.

Common causes of blindness in the world

- Trachoma
- Cataract
- Glaucoma
- Keratomalacia
- Onchocerciasis
- Diabetic retinopathy

The most common cause of irreversible blindness and partial sight in developed countries is age-related macular degeneration. In patients of working age, diabetic retinopathy is the leading cause in the West.

Colour vision and colour blindness

For normal colour vision we require cone photopigments sensitive to blue, green, and red light. The commonest hereditary colour vision defect is X-linked failure of red-green discrimination (8% ♂ and 0.5% ♀ affected—so those with Turner's syndrome have ♂ incidence and those with Klinefelter's have ♀ incidence). Blue-yellow discriminatory failure is more commonly acquired and sexes are affected equally.

Diagnosis. This is by use of coloured pattern discrimination charts (eg Ishihara plates).

Depressed colour vision may be a sensitive indicator of acquired macular or optic nerve disease.

Monochromatism This may be due to being born without cones (resulting in low visual acuity, absent colour vision, photophobia, and nystagmus), or, very rarely, due to cone monochromacy where all cones contain the same visual pigment, when there is only failure to distinguish colour.

Drugs and the eye

The eye does not retain drops for as long as ointments and two-hour applications may be needed. Eye ointments are particularly suitable for use at night and in conditions where crusting and sticking of the lids at margins occurs. Allow 5min between doses of drops to prevent overspilling.

The antibiotics most used are not those generally used systemically, fusidic acid, chloramphenicol, neomycin, gentamicin, and framycetin. All eye preparations have warnings not to use for more than one month.

Mydriatics These drugs dilate the pupil. They also cause cycloplegia and hence blur vision (warn not to drive). Pupil dilatation prior to examination is best achieved using 0.5% or 1% tropicamide which lasts for 3h. 1% cyclopentolate has an action of 24h and is preferred for producing cycloplegia for refraction of children. These drugs may be used to prevent synechiae formation in iritis. ▶In the over-60s with shallow anterior chambers (especially if a family history of glaucoma) they may precipitate acute glaucoma, so only use on ophthalmological advice.

Miotics These constrict the pupil and increase drainage of aqueous humor. They are used in the treatment of glaucoma (p 502).

Local anaesthetic Tetracaine (0.5% drops) is an example. This may be used to permit examination of a painful eye where reflex blepharospasm is a problem, and to facilitate removal of a foreign body. ▶It abolishes the corneal reflex so use to treat pain is to risk corneal damage. To relieve pain, give an eye pad, and be generous with oral analgesia.

Steroid and NSAID drops ▶Steroids are potentially dangerous as they may induce catastrophic progression of dendritic ulcers (p 480). Ophthalmoscopy may miss dendritic ulcers, and slit lamp inspection is essential. Steroid drops are being considered, eg for allergy, episcleritis, scleritis, iritis. Steroid drops ↑intraocular pressure (newer drops less so: eg rimexolone has a low IOP-elevating potential, comparable to fluorometholone and < dexamethasone & prednisolone acetate). medline:8694727 NSAID drops eg ketorolac, may obviate the need for some steroid drops. medline:103873

Iatrogenic eye disease Glaucoma may be precipitated by steroid drops (above) or by mydriatics—also tablets with anticholinergic effects (some antiparkinson drugs and tricyclic antidepressants).

A few drugs affect the retina if used chronically. Those taking ethambutol should be warned to report any visual side-effects (loss of acuity, color blindness). Chloroquine and other antimalarials are also implicated and damage may occur with long-term use, especially if high doses are used.

Obtaining and preparing antibiotic eyedrops

Fortified guttate gentamicin is 15mg/mL—the normal commercial gentamicin is 3mg/mL; penicillin 5000 units/mL, methicillin 20mg/mL, and antifungals can be obtained from the Chief Pharmacist, Moorfields Eye Hospital (tel. 020 725 33411). Antibiotics can be home-made as follows. *Gentamicin forte*: Add 2mL of 40mg/mL iv gentamicin to a 5mL bottle of commercial guttate gentamicin (3mg/mL).*

Other antibiotics can be made up using iv preparations to the required concentration using water or normal saline. These are stable for the time recommended for iv solutions in the manufacturers' Data sheets. Penicillin G can be used up to 500,000u/mL.

*From by Mr JKG Dart FRCS, Moorfields Eye Hospital, City Rd, London EC1V 2PD

Eyedrops as a cause of systemic symptoms

Drugs applied to the eye may be absorbed through the cornea and produce systemic side-effects—eg bronchospasm or bradycardia in susceptible individuals using antiglaucoma β -blocking drops, eg **bimatoprost** (Timoptol®), **carteolol** (Teoptic®), **betaxolol** (Betoptic®)—which is cardio-selective). ►Symptoms may be subtle and insidious—eg gradually decreasing exercise tolerance, or falls from arrhythmias. Serious problems are more likely if there is co-morbidity (eg respiratory infection).

Other anti-glaucoma drops (p 502) cause headaches, and a bitter taste in the mouth; urolithiasis is reported with **brinzolamide** (Trusopt®).

Pilocarpine (eg Isopto Carpine®, Sno Pilo®) may cause parasympathetic sweating, salivation, pulmonary oedema, BP↑, pulse↑, lacrimation, D&V, wheeze, and colic. Accommodation spasm may lead to brow-ache (worse if >40 years old, or just starting treatment).

Even highly-selective α_2 -receptor agonists used in glaucoma, eg **brimonidine** (Alphagan®), can cause effects such as dry mouth (in 33% of patients), headache, fatigue, and drowsiness.

medline:9230823

Contact lenses

80% of contact lenses are worn for cosmetic reasons. Only 20% are worn because lenses are more suitable for the eye condition than spectacles. Among this 20% a minority wear the lenses to hide disfiguring inoperable eye conditions, a greater proportion have them for very high refractive errors. Myopia above -12.00s and hypermetropia above $+10.00\text{s}$ are indications for lenses because equivalent spectacles produce quite distorted visual fields. Contact lenses after cataract removal come into this category. Lenses are used for ocular reasons, eg after corneal ulceration or trauma when a new front surface of the cornea is needed to see through and in keratoconus. Keratoconus is a rare degenerative, slowly progressive corneal condition with thinning and anterior protrusion of the central cornea. Blurred vision is the only symptom. Contact lens use may compensate for corneal distortion early, but later corneal grafting may be needed.

Types of lens Hard lenses are 8.5–9mm in diameter and are made of polymethylmethacrylate (PMMA). Gas-permeable hard lenses are about 0.5cm larger and are designed to allow gas to permeate through to the underlying cornea. They can only be made to cope with a limited degree of astigmatism and do not wet as well as standard hard lenses so may mist up in the day. With the advent of the larger (13–15mm diameter) soft contact lenses it was hoped that many of the problems with hard lenses could be circumvented. Soft disposable lenses can be worn for 2–4 weeks and then disposed of. Special contact lenses called toric lenses can be used to correct astigmatism of up to 2 diopters. With more astigmatism, correction is not achieved because the lens fits the astigmatic cornea taking on its shape. They are more delicate than hard lenses and need meticulous cleaning. Extended wear-lenses can be worn for up to 4 months. Sometimes coloured lenses are used simply to change eye colour. Disposable contact lenses are becoming more common.

Patients may suffer from keratoconjunctivitis or giant papillary change in the upper tarsal conjunctiva, possibly due to sensitization to the cleansing materials used or to the mucus which forms on the lens.

Cleaning lenses Different cleaning solutions made by different manufacturers should not be mixed. With hard lenses 2 solutions are usually used, one for rinsing and cleaning, and one for storage. The storage solution should be washed off before the lens is inserted. Soft contact lenses, being permeable, tend to absorb the chemicals, so weaker solutions for cleaning are used. In addition, the lenses are usually intermittently cleaned with another system (eg enzyme tablets) to remove mucoprotein on their surface. Sensitivity to cleaning agents usually presents as redness, stinging, increased lens movement, increased mucus production, and thickened eyelids. It may be necessary to stop wearing lenses for several months. When restarting use a preservative-free cleansing system.

Complications

- 1 Contact lenses are the most important risk factor for corneal ulceration. Corneal abrasion is common early while adjusting to wear. Pain \pm lacrimation occurs some hours after removing the lens.
- 2 Sensitization to cleaning agents \pm staining, eg by rifampicin or fluorescein.
- 3 Losing the lens within the eye. Hard lenses may be lost in any fornix, soft lenses are usually in upper outer fornix.
- 4 Keratitis especially for extended-wear (>6 days) soft lenses.
- 5 Risk of acanthamoeba infection.

Disposable lenses changed daily give the fewest complications, medline 11071345

Hygiene tips

Pay attention to contact lens containers, as well as lenses.

Do not assume that because a person uses disposable lenses there can be no nasty acanthamoebae infections. These free-living protozoa (found in soil and water, including bathroom tap water) may cause devastating keratitis even with disposable lenses.

www.txt.co.uk/full.cfm?0521

Scrub container's inside with cotton wool bud moistened with lens fluid.

Disinfect the container with hot water ($\geq 80^{\circ}\text{C}$); leave to dry in open air.

Wash your hands before handling the contact lens container.

Replace the container at least every year.

Protozoa may survive new '1-step' solutions of 3% hydrogen peroxide.

Amoebae are difficult to treat, and there is current interest in salicylate's potential to reduce microbial attachment when used in contact lens care solutions.

medline:10647728

Blepharospasm

Blepharospasm is involuntary contraction of orbicularis oculi. It commonly occurs in response to ocular pain. Repetitive blepharospasm, which may make the patient effectively blind, is a focal dystonia (OHCM p 342). If the condition is not recognized, it is all too easy to dismiss the patient as hysterical and to think that screwing up of the eyes is deliberate—especially as the more the sceptical doctor questions and probes the afflicted patient, the worse the blepharospasm may become (stress is an important exacerbating factor). It is important to understand that it may have a serious negative impact on patients' lives. medline:11033379

Presentation ♀:♂ ≈ 1.8:1. Blepharospasm is often preceded by exaggerated blinking. Other dystonias may be present (eg oro-mandibular). It usually starts unilaterally, becoming bilateral. Patients may develop various tricks to reduce blepharospasm such as touching or pulling the eyelids—a variation of *geste antagoniste* seen in other forms of dystonia.

Causes Mostly unknown. Recognized antecedent events are: neuroleptic drugs, Parkinson's disease, progressive supranuclear palsy.

Treatment **Drugs** Response is variable; good effects may wear off. Try anticholinergics (trihexyphenidyl, eg 1mg/day po, max 5mg/8h; tablets are 2mg or 5mg). Dopamine agonists (L-dopa, bromocriptine) may help. *Botulinum neurotoxin* Palliation is achieved with nanogram doses given to orbicularis oculi, in which it produces a temporary flaccid paralysis. It can help some people recover effective vision. It binds to peripheral nerve terminals and inhibits the release of acetylcholine. Treatment may need to be repeated, eg 3-monthly. **Supportive treatment** If the cause is compensation for apraxia of lid opening, wearing goggles may help. medline:11052590
Support is available from the UK Dystonia Society. tel: 0207 329 0797

Allergic eye disease

Allergic conjunctivitis is common, causing itch and eye watering with mild conjunctival congestion (red) and swelling (chemosis). Cold compresses may help. **Prescription drops**: levocabastine (/12h) may be better than nedocromil (/12h). medline:8977503 **Alternative**: azelastine. medline:9788686

Over-the-counter drops: cromoglicate 2% (/6h). medline:1334445

Vernal keratoconjunctivitis (spring catarrh; seasonal conjunctivitis)

This is an uncommon allergic bilateral conjunctivitis, usually appearing before puberty, and lasting ≥5yrs. ♂/♀ >1. There is often a family history of atopy. It is almost non-existent in cold areas. **Presentation**

- Itching eye ± photophobia
- Milky conjunctiva and ropy discharge
- 'Cobblestones' under upper lid
- Papillae in lower tarsal conjunctiva
- Pseudomembrane (Maxwell-Lyon)
- Corneal arcus (±ulcer) near limbus

Management If this is a very difficult problem to manage. Refer. In severe conditions, ophthalmologists will use steroid drops and taper it off. Cromoglicate (mast cell stabilizing agent) drops 2%(/6h) will be prescribed concomitantly and maintained till spring is over.

Delayed hypersensitivity: phlyctenular keratoconjunctivitis This may be a delayed response to mycobacteria, staphs, yeasts, or chlamydia. The conjunctival phlyctenule begins as a hard, red, 1–3mm diameter papule surrounded by hyperaemia. At the limbus, it is triangular (apex points to cornea). It ulcerates, and then subsides within 12 days. Corneal phlyctenules develop as an amorphous grey infiltrate, and leave a scar. They may cause intense photophobia. **Management** Seek expert help. Steroid drops may help. Treat any bacterial cause. medline:10715381

Floaters, flashes, and haloes

► When patients complain of flashes and floaters think of retinal detachment. When patients mention their haloes they may not be referring to their own saintliness. *Halo* refers to a circle of light, either white or coloured, seen around any luminous body: not just those of saints. Also call to mind Monet and his cataracts (a cause of haloes). It is not simply that his great impressionist paintings are investments in light (another definition of *halo*). . .

Doctor, you say there are no haloes caused by old age, an affliction.*
around the streetlights in Paris I tell you it has taken me all my life
and what I see is an aberration to arrive at the vision of gas lamps as angels

Here, Monet refuses surgery for his cataracts (how differently doctors and artists see the world!). So before we recommend surgery it might be as well to get to know our patients a bit, as well as keeping a weather-eye on posterity.

Floaters These are small dark spots in the visual field, particularly noticeable against a bright background. The cause is often degenerative opacities in the vitreous (syneresis), here, each floater is a shadow of a mobile vitreous opacity cast on the retina. After the eye comes to rest, they continue to move. They are common in myopes, after cataract operations or after trauma. Mostly, they are just annoying (if central), but harmless, and may settle with time. Examine the vitreous and retina before reassuring.

Sudden showers of floaters appearing in one eye (which may be accompanied by flashing lights) are due to blood. ► Refer immediately, as the cause may be retinal detachment (p 506). Other causes:

- | | | |
|-------------------------------------|--------------------|-------------|
| ● Diabetic retinopathy | ● Hypertension | ● Leukaemia |
| ● Old retinal branch vein occlusion | ● Endocarditis | ● Sarcoid |
| ● Recurrent vitreous bleeding | ● Sickle-cell dis. | ● TB; SLE |

Flashing lights (photopsia) These may be from intraocular pathology or migraine. Is there headache, nausea, or previous migraine? Detachment of a shrinking vitreous from the retina (commoner in myopes) gives flashes and floaters. 5% go on to retinal tears and detachment. Retinal damage is usually peripheral and hard to see—refer immediately for specialist help.

4 'F's of retinal detachment ● Floaters (numerous, acute onset, constant, and described as 'spider's web') ● Flashes ● Field loss (acute, progressive) ● Falling acuity. Flashes are due to pulling on the retina, and may increase with eye movement. Field loss may be seen as a dark cloud covering a field of vision (superior field loss indicates inferior retinal detachment and vice versa). Falling acuity indicates a grave situation. Superior detachments may be fast so that the macula is affected, resulting in 6/60 vision. In this situation vision remains poor even after successful surgery.

Floaters in vitreous separation ('Aggregates of collagen fibres') ● Gradual onset ● Vision unaffected ● Few in number ● No field defects ● Non-urgent.

Haloes may be from acute angle closure glaucoma. Smooth, coloured haloes around lights are diffractive phenomena, being seen when a white light is inside a steamed-up window (street lights have haloes when seen through misted windscreens), or through scratched spectacles. Hazy ocular media may also be the cause (cataract; corneal oedema).

In acute glaucoma it is corneal oedema that causes them as intraocular pressure rises with pupillary dilatation. If haloes are accompanied by eye pain consider this diagnosis and refer immediately.

Jagged haloes which change shape are usually due to migraine. Beware labelling haloes as being migrainous in patients over 50 years who have not previously suffered from migraine.

* Lisel Mueller 1986 *Monet Refuses the Operation* in *Second Language*. See M Faith McLellan 1996 (*Lancet* 1996 **348** 1641)—to whom this page owes much—for a critique of this poem.

Refractive procedures

Refractive procedures are now increasingly undertaken as an alternative to wearing spectacles; mostly for cosmetic reasons. Occasionally they are undertaken for anisometropia (imbalance of prescriptions); for astigmatism after surgery; or for intolerance of spectacles or contact lenses. A variation of PRK (photorefractive keratotomy) may be undertaken for some corneal diseases. Lasik (see below) is now the commonest procedure. It is well researched and in terms of surgical procedures is extremely safe but there are possible complications. It should be noted that the majority of ophthalmologists wear spectacles!

Radial keratotomy This is historical and should no longer be done. It involved surgical incisions of the peripheral cornea to alter the curvature.

Photorefractive keratotomy (PRK) This is an entirely laser treatment where the curvature of the front of the cornea is altered by ablation of part of it using a laser. It is only undertaken for low degrees of myopia and is less predictable than Lasik with some people having under-correction and others over-correction. Occasionally some people develop corneal haze with reduced vision, glare, and haloes afterwards. In the majority of low myopes it gives good outcome but it is very painful for a few days.

Lasik (laser assisted in situ keratomileusis) This is a combination of minor surgery where the cornea is incised and an extremely thin trapdoor shaped flap hinged away and then laser is applied to the bare corneal stroma underneath. Thereafter the flap is pushed back into position and adheres naturally. It is, surprisingly, virtually painless, settles very quickly, and is fairly predictable in its outcome. It is possible to undertake surgery on much greater degrees of refractive error; with up to 5 diopters of hypermetropia; 5 diopters of astigmatism; and 15 diopters of myopia. Serious complications are rare but trauma to or infection of the flap may result in permanent corneal scarring.

Other procedures These are much less commonly undertaken. They include insertion of perspex rings into the cornea; other laser techniques to alter corneal curvature; and surgery to the sclera to attempt to correct presbyopia. They are much less certain in outcome and are best regarded as experimental.

7. Ear, nose & throat diseases

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▶ Ideally, this chapter should be read with a ready supply of ENT patients and access to surgeons willing to teach about the various operative techniques employed. If this is impossible, a good place to start may be with a successful multimedia title such as *Otorom* (by Robert Vincent, Heinemann, ISBN 07506 3063 9).

Web-based guides to ENT examination are readily available: see
www.medinfo.ufl.edu/year1/bcs/clist/index.html

Acknowledgement: We particularly thank Mr Tor Chiu who masterminded this chapter.



A cochlear implant. An electrode is inserted into the cochlea connected to a receiver implanted in the skull. The external audio processor is attached by a magnet to the overlying skin.

ENT examination

Examination in ENT is no different from other specialties, except for the fact that the regions of interest are rather inaccessible. Head worn illumination either in the form of a head mirror with an external light source or more modern self-contained headlight is important as it provides a well-illuminated stereoscopic view of cavities, leaving the hands free. Positioning of the patient is important. Rod lens and flexible fibreoptic scopes for nasendoscopy and laryngoscopy are now part of a routine ENT examination.

Examination of the ear (Position yourself to one side of the patient—begin on the better side, if applicable). Inspect the pinna for scars (postauricular and endaural scars suggest mastoid and middle ear surgery respectively) and other abnormalities eg cauliflower ear, perichondritis.

Examine the auditory meatus by first pulling the pinna *up* and *back* to straighten the cartilaginous bend. (In infants, the bony canal has not formed fully, so pull the pinna *back* and *down*.) Swab any discharge, and remove any wax (p 530). Insert the largest comfortable aural speculum on an otoscope, taking care not to probe too deeply into the sensitive inner bony canal. Look for infection/inflammation, stenosis and exostoses (p 530).

The first thing to learn is to distinguish between a normal and an abnormal eardrum; this takes practice. Examine the quadrants in turn. Identify the pars tensa, pars flaccida, the handle of the malleus and the cone of light (light reflex) that points to the toes (ie anteroinferior). Note colour, translucency, and any bulging or retraction of the membrane. **Perforations:** Size; position; site: *marginal* or *central*? Perforations or retractions of the pars flaccida may indicate serious pathology (p 536). Assess drum mobility using a pneumatic attachment for the otoscope or a special Siegle speculum. On squeezing the balloon, the drum should move. Drum movement during a Valsalva manoeuvre means a patent Eustachian tube.

Finish with free field voice testing (whisper from 40cm) and tuning fork tests. Consider nerve VII and the postnasal space.

Examining the nose (Sit opposite the patient, with your knees together and to the patient's right). Inspect the external nose: size, shape, deviations, or deformity. Lift the tip of the nose to inspect the vestibule. Check patency of each side either with a cold metal tongue depressor or by occluding each nostril in turn with the flat of your thumb (don't press the side in: this distorts the other nostril); ask the patient to sniff. Test smell if indicated.

Thudicum speculum (anterior rhinoscopy): insert gently; assess mucosa, septum position, and the front of inferior and middle turbinates. Any polyps?

Examine the rest of the nasal cavity with rigid endoscopy after spraying with xylometazoline & lidocaine. The middle meatus is a key nasal area as most of the sinuses ventilate via this cleft. The postnasal space (nasopharynx) contains the Eustachian tube orifices & the pharyngeal recess (of Rosenmüller), and may contain adenoids or nasopharyngeal cancer. A postnasal mirror can be used to examine the nasopharynx but is only produced during clinical exams! Examine the palate, as this forms the floor of the nose.

Examining the throat Position yourself as above; ask edentulous patients to remove dentures. Inspect the lips and perioral region. First ask him to open his mouth without protruding the tongue, then use a tongue depressor to retract the buccal mucosa on each side: inspect the buccal mucosa (including parotid duct opening opposite the upper 2nd molar), gums, teeth, floor of mouth, and in particular, the retromolar trigone.

Depress the tongue; say 'ah' or 'aye' (check palate movement). Examine tonsils (prominent tonsils aren't necessarily enlarged; deep crypts with debris can be normal). Put on a glove for bimanual examination of the floor of mouth, checking the submandibular duct for stones or masses. Palpate the base of tongue for early tumours that are not easily visible.

Free field voice testing (Other tests: p 532)

- Explain what you are about to do. Stand behind the patient.
- To test the right ear, use your left hand to rub the patient's left tragus to mask that ear, then 2 ft (approximately arms length), whisper a simple polysyllable, eg 'fortyone' or 'chicken'.
- The patient should be able to repeat what is said at least 50% (or 2 out of 3) of the time; failure represents hearing impairment. This is clarified further by (a) coming closer to 6 inches and (b) raising the voice to conversational levels, it may even be necessary to shout!

Indirect laryngoscopy (This takes practice)

- Explain what you are about to do to the patient; this is very important.
- Anaesthetize the throat with a topical 2% lidocaine spray; warn the patient not to eat anything hot for 1–2h after.
- The tongue is protruded and gripped gently with a swab with slight forward tension. With the other hand warm a mirror (check it is not too hot). Ask the patient to mouth breathe, and place the mirror up to the uvula/soft palate. Examine the epiglottis and the false cords (ventricular folds). As the patient says 'ee', vocal cords and their movement may be assessed.
- Flexible nasendoscopy (4mm) can be used in those who cannot tolerate indirect laryngoscopy.

The ear

The external ear The auricle is made of fibroelastic cartilage, covered by skin. Because the tympanic membrane is set obliquely, the external auditory canal is 3cm anteriorly and 2.5cm posteriorly. The outer third is cartilaginous and has hairs and ceruminous (wax) glands in the skin. The inner two-thirds of the meatus is bony and lined with sensitive skin.

Congenital anomalies The auricle develops from 6 hillocks derived from the 1st and 2nd branchial arches that appear at 4–6 weeks with the intervening 1st branchial groove forming the external auditory canal. Any malformation may give rise to accessory tags/auricles and preauricular pit, sinus, or fistula. An infected preauricular sinus can be mistaken for an infected sebaceous cyst, but there is often a deep tract that lies close to the facial nerve. It must be removed to avoid further infection.

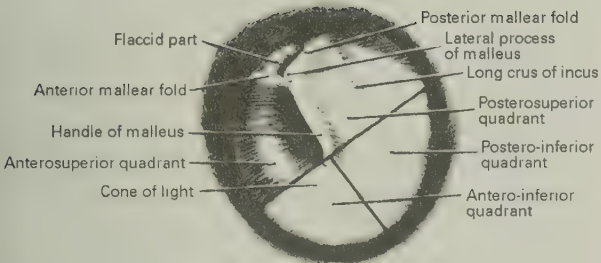
Chondrodermatitis nodularis chronica helicis This mouthful describes painful nodular lesions that usually occur on the upper margin of the pinna. It is more common in men with outdoor occupations; the exact cause is unknown, theories include sun damage or cold induced local vasoconstriction causing localized avascular chondritis. Treatment: Excision of skin and underlying cartilage (there are various methods).

Pinna haematoma Blunt trauma may cause bleeding in the subperichondrial plane that elevates the perichondrium to form a haematoma. Arrange formal, prompt evacuation. Aspiration is rarely adequate: firm packing that conforms to the contours of the auricle prevents reaccumulation. Failure to treat adequately leads to ischaemic necrosis and collapse and fibrous repair resulting in a 'cauliflower' ear.

Exostoses These are smooth multiple bilateral swellings of the bony canals and represent localized bony hypertrophy supposedly in response to cold exposure, such as in aquatic sports. Usually asymptomatic as long as the lumen is sufficient for sound conduction and thus often picked up incidentally. When they hinder migration of wax or debris, or when they occlude the canal so causing conductive deafness, surgical removal is then indicated. Osteomas are benign bony tumours that are usually solitary.

Wax (cerumen) Wax is secreted in the outer third of the canal and protects against maceration. Due to epithelial migration the ear is a self-cleaning organ; cotton buds are unnecessary and will lead to conductive deafness and discomfort if the wax impacts. Hard impacted wax may be removed by softening with oil drops (eg almond or NaHCO₃) daily for a week. Optimal treatment is suction under direct vision using a microscope. General practitioners and those without special equipment can syringe the ear with water at 37°C. The direction of the jet should pass backward and upward. Dry the ear afterwards. Avoid syringing if the tympanic membrane is perforated, or after mastoid surgery.

Foreign bodies in the ear These are common in children and the mentally retarded. Organic material may cause much inflammation. Syringing can be successful, particularly with small objects; take care with objects that almost fill the canal as these will impact and organic matter that may swell. Insects should be killed with oil before removal. Other objects require correct equipment and expertise: soft objects can be retrieved with forceps under magnification; smooth hard objects are removed with suction, otherwise they slip ever deeper into the ear. General anaesthesia may be needed in children or anxious adults.



Left tympanic membranes seen from the lateral side. The 4 arbitrary quadrants are indicated by solid lines and by the handle of the malleus. (From *Cunningham's Manual*, Vol. 3, oup.)

Hearing tests

When assessing suspected hearing loss, determine its nature (conductive or sensorineural), its severity and its cause: is it treatable, and is it part of some other disease process (eg unilateral sensorineural hearing loss (SNHL) in acoustic neuroma). Remember also to assess the disability.

Free field voice testing See p 529.

Tuning fork tests Use a 512 or 256Hz tuning fork; strike it one-third from the free end on an elbow or patella (your own) and hold it so that the 2 prongs and the meatus lie on the same line (air conduction, AC), then place the stem on the mastoid for bone conduction (BC). The patient is asked to compare loudness. Remember no single test is diagnostic but can provide useful information, and of course, are an examiner's favourite.

Rinne test $AC > BC$ = Rinne positive; this can happen with normal ears or SNHL. Rinne negative ($BC > AC$) occurs with conductive deafness more than 20dB or with severe SNHL. The latter is the classic false negative Rinne: the cochlea of the other ear picks up the sound by bone conduction. Use of Barany noise box to mask the other ear during the test, prevents this.

Webber test With the tuning fork on the vertex/forehead/upper incisors(!), ask the patient which ear the sound is heard in. Sound localizes to the affected ear with conductive deafness (>10 dB loss), to the contralateral ear in SNHL, and to the midline if both ears are normal.

Factitious deafness Usually occurs in the context of compensation claims.

Stenger test If 2 tuning forks (of equal frequency and intensity) are presented to both ears simultaneously, the sound is perceived in the midline. When one fork is brought closer the sound is heard only in that ear. If factitious deafness is suspected, ask him to close his eyes and so 'concentrate' on listening; the patient will confirm hearing a fork held 15cm away from the good ear but will deny hearing a fork held 5cm from the bad ear. Then both tuning forks, 15cm from the good ear and 5cm from the bad, are used simultaneously: with true deafness the noise is heard loudest in the good ear, but in factitious deafness it will actually be heard in the 'bad' ear but the patient will deny hearing anything, despite sound also being presented to the good ear. **Speech audiometry with feedback** As the subject reads aloud, speech is relayed back to the ear under test with a short delay. In normal hearing, this causes confusion and stammering, and a raised voice.

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Audiometry These quantify loss and determine its nature.

Pure tone audiometry is the standard measure. Headphones deliver electronically generated tones at different strengths over frequencies of 250–8000Hz in a sound-proofed room. The patient registers when he first hears the sound (the threshold) and this intensity is recorded in decibels by the tester as the air conduction threshold. A bone conduction threshold is obtained by using a transducer over the mastoid process. Masking (presenting narrow band noise to the nontest ear) is required in certain circumstances. Speech audiometry examines speech discrimination above the threshold by asking the patient to repeat the words presented through the headphones. It is useful in that it assesses disability, and can be used to predict whether a hearing aid would benefit a patient.

Both pure tone and speech audiometry are subjective tests, acoustic impedance audiometry (tympanometry) is objective. A probe with an airtight seal is introduced into the meatus; it measures the proportion of an acoustic signal reflected back at varying pressures and generates a graph of compliance. Tympanometry provides a useful measure of middle ear pressure. A normal ear shows a smooth bell-shaped compliance curve (type A). Fluid in the middle ear flattens the curve (type B). Negative middle ear pressure shifts the peak to the negative side or left (type C).

Referrals for speech therapy

For a full analysis of the problems of *delay in talking*, see p 203.

1 in 7 preschoolchildren have transient speech/language problems. Many resolve spontaneously or with professional help but serious disorders affect 1 in 20 primary and 1 in 80 secondary school children. 110,000 uk children are so seriously affected that they cannot be understood outside the immediate family. Consider speech therapy assessment if:

0–18 months of age:

- Feeding difficulties from anatomical or neurological disorder, eg cleft lip or cerebral palsy.

2 years:

- If vocabulary is <30 words or no phrases (but not if good communication skills without speech, and he or she seems on the edge of talking).

3 years:

- Speech is unintelligible.
- Using sentences of 2 words only.
- No descriptive words or pronouns used.
- Limited comprehension (eg cannot identify scissors or pen by 'which do we draw with').
- Parental anxiety, if the child is going through a stage of non-fluency.

4 years:

- Speech is not clear. (Problems with 'r', 'th' and lisps can be left until 5yrs.)
- Sentences used are less than 3 words; vocabulary is limited.
- Difficulty in carrying out simple commands.

5 years:

- Persisting articulation difficulties.
- Difficulty understanding simple sentences.
- Difficulty in giving direct answers to simple questions.
- Difficulty with sentence structure; immature sentences; word order.

6 years and older:

- Persisting articulation problem.
- Difficulty understanding spoken language.
- Difficulty with verbal expression.
- Stammering.
- Voice problems.

Children with voice problems, eg hoarseness or excessive nasality, are best referred at whatever age they present.

Painful ears (otalgia)

The cause is often non-otological (in 50%); look for sources of referred pain, eg throat and teeth (does grinding/tapping hurt—see p 572).

Otitis externa ('Swimmer's ear'): pain, discharge, and tragal tenderness due to an acute inflammation of the skin of the meatus; common predisposing factors are moisture and local trauma eg fingernails (consequently conditions causing itch eg eczema/psoriasis). *Pseudomonas* is the commonest organism involved. Aural toilet is the key to treatment.

If severe, the meatus is narrowed. A thin pope wick can be inserted and then hydrated with eardrops, eg Gentisone HC[®] (gentamicin with steroid), an alternative is using a strip of ribbon gauze soaked in glycerine-ichthammol (very soothing). After a few days, the meatus will open up enough for either microsuction or careful cleansing with cotton wool. Commercial Qtips[®] or cotton buds shouldn't be used: they are too large; instead either thin it out or make one yourself by wrapping a small piece of cotton wool gently around an orange stick. Nonspecialists may syringe the ear. Use drops only short-term, as troublesome fungal infections can arise. Beware persistent unilateral otitis externa in diabetics/ the elderly or immunosuppression: the risk is malignant/ necrotizing otitis externa that is locally aggressive and can lead to pseudomonal skull infection, for example.

Furunculosis This is an acutely painful staphylococcal abscess arising in a hair follicle. Pathologically it is identical to a boil anywhere else; if there is cellulitis consider flucloxacillin. Diabetes is an important predisposing factor.

Bullous myringitis This condition is characterized by extremely painful haemorrhagic blisters on the drum and deep meatal skin, there may also be serosanguinous fluid behind the drum. Classically associated with influenza infection, but *Mycoplasma pneumoniae* has also been implicated as having a variety of other organisms. It may simply represent a variant of acute otitis media. Sensorineural hearing loss is much more frequent than previously thought. Treatment is generally supportive only; oral antibiotics can be considered in cases with middle ear effusions.

Barotrauma (aerotitis) When the Eustachian tube is occluded, middle ear pressure cannot be equalized, during descent in an aircraft or diving so causing damage. Conditions that inhibit function of the eustachian tube eg inflammation/infection will predispose to barotrauma. There is severe pain as the drum becomes indrawn; transudation or frank bleeding into middle ear. Barotrauma to the inner ear causes vertigo, tinnitus, and deafness.

Prevention Avoid flying with a URTI; try decongestants (eg xylometazoline every 20min) into the nose or repeated yawns, swallows, and jaw movements—and Valsalva manoeuvres during descent. In one study, Valsalvas normalized pressures in 46% of adults (33% of the children). Most of these with unsuccessful Valsalvas could improve middle ear pressure by inflating an Otovent[®] device, which has been recommended to air passengers with problems clearing the ears during flight.

medline:9451533

With simple barotrauma, treatment is supportive; effusions usually clear spontaneously, and most perforations heal.

www.clinicalevidence.org

Temporomandibular joint dysfunction See p 695. Earache, facial pain, and joint clicking/popping related to malocclusion, teeth-grinding or joint derangement, and, importantly, stress—making this a paradigm of a biopsychosocial disorder which may become a chronic pain syndrome (p 695). medline:10635197

Joint tenderness exacerbated by lateral movement of the open jaw, or trigger points in the pterygoids. **Treatment** NSAIDs; cognitive therapy (p 370); physiotherapy; biofeedback (meta-analysis evidence). medline 10425966 & 10332343

Referred otalgia: when the cause is not in the ear

- Referred (secondary) pain can arise from disease processes in the territories of the sensory nerves supplying the ear.
- The great auricular nerve C2,3 (supplies lower half of the pinna) may refer pain from soft tissue injury in the neck and from cervical spondylosis/arthritis.
- The auriculotemporal nerve (trigeminal) (supplies lateral lower half of pinna) may refer pain from dental disease and TMJ dysfunction (temporomandibular joint dysfunction, opposite).
- The tympanic branch of the glossopharyngeal nerve and the auricular branch of the vagus (supplies medial surface of drum) refer pain from the carcinoma of the posterior third of the tongue, pyriform fossa, or larynx, or from the throat to the ear, eg in tonsillitis; quinsy. Otagia is common post-tonsillectomy, particularly in adults. It is worth warning all patients.
- A sensory branch of the facial nerve (supplies lateral surface of drum) refers pain in geniculate herpes (Ramsay Hunt syndrome, p 756).
- Primary glossopharyngeal neuralgia is a rare cause of pain often induced by talking or swallowing.

Discharging ears

The character of the discharge provides clues to its source and cause:

- **External ear:** Inflammation, ie otitis externa (OE) produces a watery discharge as there are no mucinous glands. Blood can result from trauma to the canal. Liquid wax can sometimes 'leak' out. ► Beware persistent unilateral otitis externa in those with diabetes or immunosuppression: the risk is *necrotizing otitis externa* (eg with *Pseudomonas* skull infection).
- **Middle ear:** Mucous discharges are almost always due to middle ear disease. Serosanguinous discharge suggests a granular mucosa of chronic otitis media (COM). An offensive discharge suggests cholesteatoma.
- **CSF otorrhoea:** CSF leaks may follow trauma or surgery. Clues to the identification of CSF include the halo sign on filter paper, or measuring glucose levels or by the presence of β_2 -transferrin.

Common causes: otitis externa, acute otitis media (AOM)—but always be wary of cholesteatoma. NB: crust in the attic (pars flaccida/posterosuperior quadrant) often hides pathology, so remove it.

Otitis media is defined as inflammation in the middle ear and classification is mostly usefully based on the timing: acute, subacute, and chronic. Acute OM presents with rapid onset of pain and fever, and sometimes with irritability, anorexia, or vomiting, often after a viral upper respiratory infection. Chronic OM (serous, secretory, or glue ear) is inflammation with middle ear fluid of several months duration.

In AOM, drum bulging causes pain, and may be followed by a profuse purulent discharge as the drum perforates. Discharge usually settles after 48h. Give analgesia \pm antibiotics (eg amoxicillin; common organisms are *Pneumococcus*, *Haemophilus*, other streps and staphs). In children, use amoxicillin in preference to penicillin as *Haemophilus* is commoner. Decongestants have little rôle in AOM.

Continuing discharge may indicate complications eg mastoiditis which is rare, and even rarer are petrositis; labyrinthitis; facial palsy; meningitis; intracranial abscesses. Mucopus may continue to drain when there is no mastoiditis, especially if grommets are in place. Treat with appropriate oral antibiotic according to swab results. Do aural toilet to remove infected material from meatus. If discharge continues get expert help.

Chronic suppurative otitis media This presents with discharge, hearing loss, but no pain. Central perforations indicate disease in the lower anterior part of the middle ear. Treatment aims to dry up the discharge (aural toilet, antibiotic with steroid eardrops, depending on culture) and surgery to correct ossicle discontinuities and to close the drum.

Cholesteatoma is serious—and a serious misnomer as it is neither a tumour nor made of cholesterol. It is skin (stratified squamous epithelium) in the middle ear that is locally destructive, through a combination of cholesteatoma growth and collagenases. **Classification.** Congenital (classically occur with an intact eardrum) or acquired, which are much commoner and can be primary (accumulation within a retraction pocket) or secondary (ingrowth of skin through a perforation). It has a distinctive pearly white appearance with surrounding inflammation.

Diagnosis The patient may complain of foul-discharge or deafness, but often the patient does not present until later with headache, pain, facial paralysis, and vertigo, indicating impending intracranial complications such as meningitis and cerebral abscess. **Treatment** Surgery is inevitable and aims to first provide a safe dry ear by removing the disease, improving or maintaining hearing is secondary.

Use of eardrops in otitis externa

► Take a swab first, and perform aural toilet.

- Betamethasone 0.1% (eg Betnesol®)—very cheap and good for non-infected, eczematous otitis externa.
- Betamethasone 0.1% + neomycin 0.5% (Betnesol-N®)—use if infected. NB: *prolonged use of steroid drops may cause fungal otitis.*
- Oxytetracycline 0.5% with polymyxin B 0.12% and hydrocortisone (Terra-Cortril® drops)—good if there is mixed infection, including anaerobes (3 times the price of Betnesol-N®)
- Gentamicin 0.3% with hydrocortisone 1% (eg Gentisone HC® drops; 6 times the price of Betnesol® or Betnesol-N®) is good, but is recommended by some only as a 2nd-line agent, eg in *Pseudomonas* infections.
- Clioquinol 1% + flumethasone 0.02% (Locorten-Vioform®) or clotrimazole topical 1% solution are used in fungal infections.
- Sodium bicarbonate 5% drops are useful if wax is an additional problem.

Fluid in the middle ear

This is detected by otoscopy (fluid level or bubbles behind the drum) or indirectly, by tympanometry. 50% of 3yr-olds have ≥ 1 ear effusion/year.¹

Glue ear Synonyms: otitis media with effusion (OME); serous otitis media.

►Hearing impairment noticed by parents is the mode of presentation in ~80%.

The fundamental problem lies with dysfunction of the Eustachian tubes (ET). Although the exact cause is unclear there are associations with upper respiratory tract infections, oversized adenoids and narrow nasopharyngeal dimensions. Large adenoids themselves do not cause OME but are important because, if infected, they are a source of pathogenic bacteria.

Glue ear is the commonest cause of hearing loss in young children, and can cause disastrous impairment in learning, though this is rare. Glue ear may cause no pain, and so its presence may not be suspected. It is commoner in: boys, atopsics, Down's syndrome, and in those with cleft palates. It is also more prevalent in winter and in the children of smoking parents.

Signs Variable. The drum can be retracted or bulging. It can look dull, grey, or yellow. There may be bubbles or a fluid level, there may be superficial radial vessels. Drum mobility when tested with a Siegle speculum or a pneumatic attachment is reduced.

Tests. Audiograms (look for conductive defects)

Impedance audiometry (look for flat tympanogram; type b)—this helps distinguish glue ear from simple Eustachian tube dysfunction and otosclerosis.

Microbiology: In 33%, bacteria can be cultured from the middle ear.

Treatment: The natural history of the condition tends towards spontaneous resolution. Simple explanation and reassurance with a 3-monthly review may be enough. Drugs have little rôle: neither decongestants nor mucolytics improve glue ear. Tonsillectomy does not help either.² Some favour long-term low dose antibiotics but the data is questionable.³

If fluid persists and hearing is impaired, consider myringotomy with suction of fluid, and insertion of air-conducting grommets* \pm adenoidectomy (so reducing recurrences but adding a small extra mortality risk). Grommets are preferred to T-tubes, which are associated with an unacceptably high rate of residual perforation (50%). The main complications of grommets are infections and tympanosclerosis. Treat infections with aural toilet and antibiotic/steroid ear drops, but removal of the grommet may be required. Autoinflation of the Eustachian tube using Otovent® balloon devices has been shown to be useful but are only effective during the period of use, and while they are less effective than grommets, they may be sufficient until time takes over.

90,000 operations/yr are done for glue ear in the UK (1 in 200 children).³

Aftercare: It is all right to swim after grommet insertion, but avoid forcing water into the middle ear by diving. Though some form of ear plug is probably prudent especially when shampooing (because soap reduces the surface tension of the water). Grommets extrude after a variable period of 3–12 months, and a review is required to recheck the hearing. A significant proportion (25%) will require further subsequent grommet re-insertion.

►In adults, exclude a postnasal space tumour as the cause of middle ear fluid.

Prognosis: Language development sequelae, eg \downarrow reading skills & IQ \downarrow , can persist into late childhood and the early teens.

2001 medline:11466180

*Benefits will be greatest in those having an effusive ear for >90 days with a hearing loss of >20 dB. 1 K Bennett 1999 Arch Dis Child 80 28 2 J Counel 1995 Lancet 345 3 3 F van Balen 1996 Lancet 348 713 & EBM 1997 41

Who should be referred with glue ear?

Refer to community audiology service if not able to:

- Determination of severity of hearing loss.
- Characterization of hearing loss. Is it conductive?
- Is the hearing loss leading to a disability?

Refer direct to an ENT consultant if:

- Deaf for over 3 months—or
- Difficulties with speech or learning—or
- Recurrent and persisting earache over a 3-month period—or
- Other disabilities making correction of deafness urgent.

► NB: explain that immediate surgery is rarely justified.

Possibilities while the patient is waiting to be seen:

- Discourage passive smoking. What's it like at the childminder's?
- Encourage drinking from a cup, not a bottle (better Eustachian function).
- Encourage interlocutors to sit at the child's level, and to keep instructions short and simple. Explain the problem to all.
- Turn off the tv (less background noise, and this makes it more likely that the patient will look at the person speaking to him).
- Tell the teacher which side hears best (for class seating plan).
- Nasal steroids for 1 month—or
- Co-amoxiclav for 14 days (may be of little real benefit)—or
- Oral steroids for 5 days (eg prednisolone 1mg/kg/day po).

NB: benefit may not be long-lasting, but medical treatment may diminish the need for surgery.

Modified West Sussex protocol

Childhood deafness

Subdivisions • *Hereditary vs acquired*: Hereditary defects may be present at birth (congenital) or appear later. • *Conductive or sensorineural hearing loss: SNHL or mixed* • *Isolated deafness vs syndrome complex*.

Autosomal dominant syndromes Waardenburg's (SNHL, pigment abnormalities & telecanthus, ie ↑ distance between medial canthi) Klippel–Feil (p 752); Treacher–Collins' (p 758); Pierre Robin (p 754) Alport's syndrome (p 742).

Autosomal recessive syndromes Pendred (SNHL with goitre); Usher's (SNHL, retinitis pigmentosa); Jewell–Lange–Nielson (SNHL, long QT interval).

Acquired *In utero*: Maternal infection (rubella, CMV, influenza, syphilis) ototoxic drugs and metabolic upset eg maternal diabetes.

Perinatal: Anoxia, birth trauma, cerebral palsy, kernicterus.

Postnatal: Meningitis, chiefly; also ototoxic drugs, lead, skull fracture.

Universal newborn hearing screening (UNHS) Screening within hours of birth is the best way to ensure that deafness is diagnosed and management implemented *before* 6 months of age. ► *The aim is to avoid language delay*. This is best achieved by detecting *otoacoustic emissions* (OAE; a microphone in the external meatus detects tiny cochlear sounds produced by movement in its basilar membrane). This is abnormal or equivocal in 3–8% of neonates.

medline:10193119

Most of these 'failures' (84%) have external ear canal obstruction (collapsed ear canal or debris). medline:10529905 In these *audiological brainstem responses* (ABR) should be measured.

medline:11283492 & 11261458

Prevalence of deafness found at UNHS 0.9–3.24/1000 for permanent bilateral hearing loss of >35dB; 5.95/1000 when unilateral and moderate hearing loss infants are counted.

medline:10079793

Note that no test performs perfectly.

medline:11059707

Subjective hearing tests in the older child

- 7 months: *Directed testing* (suitable if aged 6–18 months): As the child is sat on parent's lap, an assistant in front attracts the child's attention while a tester attempts to distract by making noises behind and beside child, eg with a rattle, conversational voice.
- 2–4 years: *Distraction testing* or *conditioned response audiometry* (at 24–60 months): the child is trained to put pegs into holes or give toys to a parent, on a particular auditory cue. This method can also obtain PTAs (pure tone audiograms) in an older child. *Speech discrimination* (24–60 months): the child touches selected objects cued by acoustically similar phrases, eg key/tree.
- 5 years: pure tone audiogram.

Treatment Once deafness has been detected treatment aims to provide as good hearing as possible to help speaking and education. Teachers of the deaf make arrangements for fitting hearing aids and help monitor progress. Children usually need higher gain from their hearing aids than adults. Ear moulds may need frequent changing to maintain a good fit. Encourage parents to talk as much as possible to their deaf children. Children may be educated at ordinary schools with visits from teachers of the deaf, or, for the partially hearing, in special units in ordinary schools, or in schools for the deaf, depending on need. For some, a cochlear implant may be suitable (p 542 and OPPOSITE). NB: The shorter the duration of deafness, the better the outcome: so funding decisions should not be delayed.

Safety issues with cochlear implants

They may be damaged by direct trauma, MRI, surgical diathermy, dental pulp testers and 'therapeutic' (short wave and microwave) diathermy used in physiotherapy departments. See www.bcig.org telephone 0207 915 301

Ethical issues surrounding cochlear implants

These relate to adults and children, and revolve around cost, consent, and quality of life—and *who* should decide. The situation is complex as technical improvements ensure an ever-changing balance between benefits and risks (rejection of the device for difficult-to-predict psychological reasons, and post-op complications, eg VII palsy, and stimulator migration)—so beware simplistic statements coming from meta-analysisists making statements such as 'cochlear implants are suitable for such-and-such a group (eg profoundly deaf adults) but not another group (eg congenital deafness)'. Also beware of statements such as 'after cochlear implant patients can lead normal lives' (this may only mean that *other people* don't notice the hearing problem). Post-op training is vital, and the user is by no means an inactive recipient. Remember that society is often biased in favour of technology (the black box that does everything), while all too often neglecting the human processes of communication.

The wealth of a society can be gauged by the level of deafness at which most cochlear implants are done. In the UK, cochlear implant candidates generally have no hearing (>110dB loss). There are ~8000 such candidates in the UK (under the age of 70). If the criterion is 'relaxed' to 105dB, this number rises to 45,000. In the USA, the loss is generally >95dB (in the UK this would lead to 72,000 candidates and a bill of £2520 million).

For multichannel cochlear implants, the average cost per QALY (quality-adjusted life-year) is ~£11,400/QALY² which is roughly in the middle of quoted estimates for costed NHS therapies (for a list, see *OHCM* p 14). On this view, implantation of those at least up to 70 years old makes good sense in terms of health economics. Furthermore, multiple disabilities are no bar to getting benefit from a device, but it is not clear that they merit extra consideration (they certainly merit very careful assessment).

Who should decide these issues is unclear (as usual!), but deaf people early should have some input (in the USA, no deaf people were on the board which decided these issues).

Cost per QALY may fall with further technical advances—eg beam-forming dual microphone systems to aid hearing in background noise, better speech processors, and new electrode arrays where a greater number of electrodes lie close to the spiral ganglion cells.³ Difficulties in assessment are compounded by the need not simply to take into account hearing and language, but also learning, socialization, and other cultural variables.

For a diagram about cochlear implants, see p 527.

DARE Meta-analyses 1997 *The Cochlear Implant in Adults, Adolescents, and Children*. 2 G Donoghue 1996 *Health Trends* 28 106 3 G Clark 1998 *Auris Nasus Larynx* 25 73

Deafness in adults

3 million adults are hearing-impaired (UK). It can be a frustrating and isolating disability. A life without intonation can be humourless—but many people cope well with some degree of hearing loss.

Management Classify the deafness (OPPOSITE); find treatable cause. Exclude the dangerous (particularly the unilateral/asymmetric): acoustic neuroma, cholesteatoma, effusion from nasopharyngeal carcinoma. *Sudden sensorineural deafness demands immediate specialist referral.*

Definitive management may be required, eg surgery for perforations or otosclerosis—or simple fitting of the most suitable type of hearing aid.

Those with profound sensorineural deafness (>100 – 120 dB) may benefit from a cochlear implant (diagram, p 527) to stimulate residual neural tissue; it takes ~2h to fit (under GA); an external device processes sound and transmits it across the skin to the subcutaneous receiver coil—to an electrode placed within the cochlea via the round window and directly stimulates the auditory nerve. The signal is not normal sound; rehabilitation needed to understand the new sounds. Benefits include improved lip-reading and recognition of environmental sounds, and relieving isolation. Cost (UK) up to £35,000 per procedure. Good pre-op assessment is vital. Selection criteria include cause (exclude central deafness), timing (most useful in postlingual patients), age (no limit) and health, commitment etc.

Sudden hearing loss (HL) Conductive causes: infective, occlusive, trauma, and iatrogenic sensorineural causes: iatrogenic, fractures.

One finds a diagnosis in nearly all cases of sudden conductive HL but up to 60% of sensorineural HL are labelled idiopathic; 60% of these will recover spontaneously. All cases of sudden sensorineural HL require urgent referral for assessment, monitoring (stable or progressive) and special investigations/treatment (viral titres, evoked response audiometry \pm MRI) and rehabilitation. Treatment is controversial and empirical, but most agree that if treatment is instituted then it should be given early, eg within 48h.

Otosclerosis Prevalence 0.5–2% clinically and 10% subclinically, although this is disputed. Autosomal dominant with incomplete penetrance. 50% have a family history. 85% bilateral with a female/male ratio of 2:1.

Vascular spongy bone replaces normal lamellar bone of otic capsule origin particularly around the oval window which fixes the stapes footplate. Symptoms usually appear in early adult life and are made worse by pregnancy, menstruation and the menopause. There is conductive deafness (hearing is often better with background noise), 75% have tinnitus and mild, transient vertigo is common too. 10% have Schwartz's sign—pink tinge to the tympanic membrane; audiometry with masked bone conduction shows a dip at 2kHz (Cahart's notch). Fluoride (controversial), hearing aid or surgery (stapedectomy/stapedotomy) replacing the adherent stapes with an implant helps 90%. Many prefer surgery to wearing a hearing aid and there is a 1–4% risk of a dead ear.

Presbycusis (senile deafness) Loss of acuity for high-frequency sound starts before 30yrs old and rate of loss for the higher frequencies is progressive thereafter. Deafness (reflecting loss of hair cells) is gradual in onset and we do not usually notice it until hearing of speech is affected. This may occur with loss of high-frequency sounds (consonants at ~3–4Hz are needed for speech discrimination). Hearing is most affected in the presence of background noise. Hearing aids are the usual treatment.

Classification of deafness

Conductive deafness There is impaired sound transmission via the external canal and middle ear ossicles to the foot of the stapes through a variety of causes. External canal obstruction (impacted wax, discharge from otitis externa, foreign body, developmental abnormalities); drum perforation (trauma, barotrauma, infection); problems with the ossicular chain (otosclerosis, infection, trauma); and inadequate Eustachian tube ventilation of the middle ear with effusion present (eg secondary to nasopharyngeal carcinoma) all result in conductive deafness.

Sensorineural deafness This results from defects central to the oval window in the cochlea (sensory), cochlear nerve (neural) or, rarely, more central pathways. Ototoxic drugs, eg streptomycin and aminoglycosides (esp. gentamicin), postinfective (meningitis, measles, mumps, flu, herpes, syphilis), cochlear vascular disease, Ménière's (p 546), trauma, and presbycusis are all sensorineural. Rare causes: acoustic neuroma, B₁₂ deficiency, multiple sclerosis, secondary carcinoma in the brain.

Tips on communicating with those who are hard of hearing

- Decrease background noise.
- Use short sentences.
- Place yourself on the same level as the person you are speaking with.
- Ensure that light falls on your face.

Tinnitus

In the ears of my days
Is a thunder of accomplished rivers e.e cummings

Tinnitus (Latin *tinnire* meaning to ring) is a sensation of sound not due to external stimuli. It is usually ringing, buzzing, or whooshing in the ears. It excludes bruits and other vascular noises. Transient tinnitus is common but only 0.5–2% are severely affected. Peak age of onset is 50–60 years.

The mechanism This is obscure. It may represent:

- Spontaneous otoacoustic emission.
- Cross-talk between adjacent nerve fibres from myelin damage after trauma.
- Altered central processing. This is being recognized as more important and helps explain the psychological associations.

Causes Often unknown—but may be related to almost any ear disease. **Some:** Hearing loss (20%); presbycusis; noise-induced; head injury; otosclerosis/post-stapedectomy; Ménière's. **Systemic:** Cardiovascular: BP↑, Hb↓ heart failure. **Drugs:** Aspirin; loop diuretics; aminoglycosides, quinine, alcohol excess. **Psychological associations:** Redundancy, divorce, retirement.

A holistic approach to tinnitus entails: time spent on the history; doing tests to exclude transmitted noise (eg glomus jugulare tumours); treating the treatable causes—and to offer support to help ameliorate symptoms.

History Site; character (constant? pulsatile?); alleviating and exacerbating factors; otalgia; otorrhoea; vertigo; head injury; family history of deafness or tinnitus; social surroundings (tinnitus is worse if isolated or depressed); drugs. Assess the disability, and, in particular, if it disturbs sleep.

Examination Otoscopy to detect tympanic membrane mobility (patulous Eustachian tube or myoclonus of tensor tympani) and middle ear disease; hearing tests including tuning fork. Check BP and pulse.

Tests Audiometry (with masking), tympanogram (including stapedial reflex thresholds). ▶ Investigate unilateral tinnitus fully (eg MRI or brainstem evoked response audiometry, ERA) to exclude an acoustic neuroma (p 562; 10% present this way). Others: Hb; syphilis serology, lipids.

Treatment Once serious disease is excluded, often simple reassurance alone is enough. A positive attitude is important; do not be dismissive and one should avoid terms such as 'untreatable' or 'you must learn to live with it'. Treat the person as a whole, not just as a malfunctioning ear.

Deafness If there is significant deafness (>35dB) a hearing aid that improves the perception of background noise makes tinnitus less apparent. A masking device generating white noise can be worn invisibly in the canal; a portable radio tuned 'off station' under the pillow can aid sleep.

Psychological support is central to relieving tinnitus. Hearing therapists can employ tinnitus restraining therapy to make tinnitus less intrusive, partly by dealing with inappropriate beliefs and fears, and partly by devices that provide a low level of background sound, which may gradually decrease CNS oversensitivity to auditory neural signals. Cognitive therapy (p 370) also helps (meta-analyses support this¹), as do patient support groups.²

Drugs Avoid tranquillizers, particularly if the patient is depressed (nortryptiline is best here), but hypnotics at night may help. Carbamazepine is disappointing; betahistine only helps if Ménière's is the cause. Randomized trials support use of melatonin (3mg nocte)³ and baclofen (10mg/12h po increasing to 30mg/12h over 3 weeks).⁴ These latter two are non-standard.

Vertigo

Vertigo (the illusion of movement) can be severely disabling. Look for secondary complaints of feeling unreal, or panic, or loss of memory. Find out if it lasts seconds (usually benign positional vertigo, below, if no CNS signs), hours (eg migraine), or days (look for a central cause, ie of nerve VII, brainstem vestibular nuclei, medial longitudinal fasciculus, cerebellum, or vestibulospinal tract—see below).

Vestibular (peripheral) vertigo is often severe, and may be accompanied by nausea, vomiting, hearing loss, tinnitus, and nystagmus (usually horizontal). Hearing loss and tinnitus are less common in central vertigo (it is usually less severe). Nystagmus may be horizontal or vertical with central vertigo—and may be different in each eye (eg in the abducting eye).

Causes Peripheral:

Ménière's disease

Benign positional (postural) vertigo (BPV)

Vestibular failure/insufficiency; labyrinthitis

Central:

Acoustic neuroma

Multiple sclerosis

Head injury; inner ear syphilis

Vertebrobasilar insufficiency

Drugs eg gentamicin (neuronitis), diuretics, cotrimoxazole, metronidazole. Effects may be central or ototoxic, **Mass lesions** Cholesteatoma; trauma.

History Although patients will often find it difficult to describe their symptoms, it is important to distinguish vestibular from non-vestibular symptoms such as lightheadedness and dizziness which can result from cardiovascular, ocular, musculoskeletal, and metabolic causes as well as claustrophobia. Associated factors such as a history of traumatic and the time scale/duration of symptoms provides important clues.

Examination and tests Must include a full neurological and otological (especially middle ear) assessment. Test cranial nerves, cerebellar function and reflexes. Assess nystagmus. Assess gait, and Romberg's test (+ve if balance is worse when eyes are shut—implying defective joint position sense or vestibular input) and Unterbergers (ask patient to march up and down on the spot with arms stretched out in front and eyes closed. Do provocation tests (Hallpike, below). If equivocal, consider audiometry; electronystagmography; brainstem auditory evoked responses; calorimetry (the only way to test each labyrinth separately; irrigate each canal with water 7°C above and 7°C below body temperature—is nystagmus induced?); CT; MRI; EEG; LP.

Benign positional vertigo (BPV) Attacks of sudden-onset rotational vertigo lasting >30sec provoked by head-turning. Common after head injury. Other otological symptoms are rare. **Pathogenesis** Displacement of otoconia in the semicircular canals. **Causes** Head injury; spontaneous degeneration of the labyrinth, post-viral illness, or stapes surgery; chronic middle-ear disease.

Diagnosis Hallpike test: This manoeuvre rotates the posterior semicircular canal in the plane of gravity. Explain and reassure. Ask the patient to keep their eyes open and to look straight ahead. With the patient supine, the head is held between the examiner's hands, turned 30–40° to one side and then rapidly lowered ~30° below the level of the couch. Ask the patient if they feel dizzy and look for nystagmus. If +ve, the patient experiences vertigo and rotary nystagmus towards the undermost ear, after a latent period of 5–10sec. This lasts <1min (adaption). On sitting, there is more vertigo (± nystagmus). If any of these features are absent (ie no latency, no symptoms, and persistent nystagmus), seek a central cause.

www.arom.com/vestib/glossary.html#H

Usually self-limiting within months; if persistent, try physiotherapy referral for teaching vestibular exercises for rehabilitation (habituation). **Counselling and reassurance** is most important; habituation (ie repeated adop-

on of the position causing vertigo) is the mainstay. ↓Alcohol intake may help. *Medication:* histamine analogues (betahistine), vestibular sedatives (prochlorperazine) or antidepressants. *Last resort:* posterior semicircular canal neurectomy or obliteration by transmastoid laser, but deafness may follow.

Ménière's disease Dilatation of the endolymphatic spaces of the membranous labyrinth causes vertigo for up to 12h with prostration, nausea and vomiting; uni- or bilateral tinnitus occurs ± progressive sensorineural deafness. Attacks occur in clusters. Treat acute vertigo symptomatically (cyclizine 50mg/8h po). Betahistine 16mg/8h po is unpredictable but is worth trying; diuretics (chlorthalidone) probably less so. Operative decompression of the saccus endolymphaticus may relieve vertigo, prevent progress of the disease, and conserve hearing. Vestibular neurectomy/labyrinthectomy may relieve vertigo but causes total ipsilateral deafness. Day-case transtympanic instillation of ototoxic drugs (gentamicin via a drummet) can help, and usually avoids deafness.

Vestibular neuronitis follows a febrile illness in adults, eg in winter, and is probably viral. Sudden vertigo, vomiting, and prostration are exacerbated by head movement. Treatment: try cyclizine 50mg/8h po. Improvement occurs in days, though full recovery occurs within 2–3 weeks. It is difficult to distinguish from 'viral labyrinthitis'.

Chronic nasal obstruction

Causes *Child* Large adenoids, rhinitis, choanal atresia, postnasal space tumours (eg angiofibromata), foreign body (►refer same day—eg if **unilateral** obstruction ± foul or bloody discharge). *Adult* Deflected nasal septum, rhinitis, polyps, sinusitis, granuloma (TB, syphilis, Wegener's, leprosy), topical vasoconstrictors, tricyclics. ►Refer urgently if: •Airway blocked by enlarged turbinate •Unilateral obstructing mass •A tumour may be present.

Diagnosis Ask about variability of symptoms, the pattern of obstruction and effects on eating, speech, and sleep (snoring). Examine to assess any nasal deflection. Is either nostril *completely* blocked (hold a mirror under each nostril: does it steam up)? Examine the postnasal space with a mirror (in children imaging the postnasal space with lateral radiographs is easier).

Rhinitis medicamentosa Drops or sprays which decongest by vasoconstriction can damage the mucosa, due to hypoxia, with rebound engorgement and mucosal oedema encouraging further drug usage. The mucosa is swollen and red. Such decongestants should not be used for >one week.

Vasomotor rhinitis This causes **variable, bilateral** obstruction ± rhinorrhoea. Swollen oedematous turbinates may be seen and there is excess mucus. **Treatment.** Measures as for allergic rhinitis are usually ineffective. Ipratropium nasal spray 2 × 20µg puffs/nostril/6h helps rhinorrhoea. Cautery or surgical reduction of inferior turbinates helps 'blocked nose'.

Deviated nasal septum Rare in children, this may affect 20% of adults. It may be secondary to injury. Surgical filleting of the bone and cartilage of the nasal septum—septoplasty repositions the septum into the midline.

Allergic rhinitis may be **seasonal** (hay fever, prevalence ≈ 2%, high risk at 5–14 yrs old) or **perennial**. **Cause.** Eg house dust mite; pollen. **Symptoms:** Sneezing; pruritus; rhinorrhoea (eg **bilateral & variable**). **Signs:** Turbinates may be swollen and the mucosa pale or mauve; nasal polyps. Skin tests may show allergens (avoid these if there is eczema, past allergic reactions, dermatographism, or on interfering antihistamines or steroids). Here, consider radio-allergosorbent (RAST) tests to find specific IgE. **Rx:** Desensitizing injections may help ≤70% with seasonal allergy, but only 50% of those allergic to house dust mite. They may cause anaphylaxis so watch for >1h post-injection. Have resuscitation facilities to hand. **Other drugs:** Antihistamines (OPPOSITE); systemic decongestants (pseudoephedrine 60mg/12h po; CI: BP↑, hyperthyroidism, angina, myocardial infarction, MAOIs use), nasal sprays (eg sodium cromoglycate 2%, 2 × 2.6mg squeezes/4–6h), or nasal steroids, eg beclometasone 8 × 50µg puffs/24h. Steroid puffers may be used indefinitely, but steroid drops are systemically absorbed so should be used for <1 month at a time, <6 times/year. Oral steroids or stat depot triamcinolone 40–100mg IM (buttock) may tide hay fever patients over difficult times. **Herbal remedies** (bog rhubarb, ie butterbur/*Petasites hybridus*, containing pestasines) are controversial.

medline 11799030

Nasal obstruction with chronic sinusitis Give steroid drops as above for 1 month, with intermittent antibiotics when nasal discharge is purulent. Refer if there is no response to this. (Radiographic imaging is unreliable.)

Other causes of chronic rhinorrhoea & rhinitis Allergies, foreign body, CSF (eg after head injury; +ve for glucose), bacteria (including TB), AIDS, cystic fibrosis, Kartagener's syndrome, rheumatoid arthritis, sarcoid, SLE, Wegener's or other granulomata (OHCM p 736), atrophic, old age (old man's drip), pregnancy, the Pill, β-blockers, NSAIDs, overuse of decongestants, antibody deficiency (p 201), non-allergic rhinitis with eosinophilia (NARES).

Reducing mite load to help allergic rhinitis

Many strategies are tried; 3 have proved effective in (rather small) randomized trials. Their effect on symptoms is not guaranteed.

- Agents that destroy mites (acaricides)
- High-efficiency particulate air filters (HEPA)
- Bedroom-based environmental control programmes

Cochrane
medline:11687117

Which antihistamine is best?

Cetirizine, ^{£8.73} desloratadine, ^{£7.57} fexofenadine, ^{£7.40} and mizolastine ^{£6.20} are all similar at a one-tablet-a-day dose and choice may be according to patient preference, availability, interactions, and price. Typically, antihistamines interact eg with erythromycin and (other macrolides), grapefruit juice, fluoxetine, and systemic ketoconazole, but these effects are not seen with desloratadine. Antihistamines (except desloratadine) may cause ↑QR interval ± arrhythmias.

medline:11424898

When the above have been compared in pollen-induced rhinitis, they were all better than placebo, and had similar effects *vis à vis* nasal smears assessing eosinophilia, histamine/grass pollen skin tests, and grass pollen nasal provocation tests.

medline:11906367

Nasal polyps

The typical patient is a man over 40yrs old; in children they are generally associated with cystic fibrosis.

Histology: ~90% are oedematous eosinophilic polyps. Typical sites: middle turbinates; middle meatus; ethmoids. Single, benign maxillary polyps may arise in the maxillary antrum, and prolapse into the nose, and fill the nasopharynx (antrochoanal polyps).

Associations: Allergic rhinitis, chronic ethmoid sinusitis, cystic fibrosis.

Symptoms: Watery anterior rhinorrhoea, purulent postnasal drip, nasal obstruction, change in voice quality, anosmia and taste disturbance, mucocele (presents with a lump on the medial superior border of the orbit).

Signs: It can be hard to distinguish hypertrophied inferior turbinates from nasal polyps—but gentle palpation will show that the polyp, if that is what it is, is insensitive, pale, and mobile.

Drugs: Try 2 days of betamethasone sodium phosphate 0.1% nasal drops, instilled every 8h (longer courses may be safe). ► *Use of nose drops requires the correct posture:* bent double at the hips, with nostrils pointing at the sky—eg kneeling with the forehead on the floor. This may make polyps quickly diminish in size, and maintenance therapy with beclometasone spray may then be tried, eg 8 puffs/24h (50µg/puff).

Surgery: Polypectomy, eg by endoscopy. Local anaesthesia may have the advantage of a more bloodless field, along with patient awareness, should the orbit or intracranial fossa be breeched. If more than simple polypectomy is planned, it is wise to do CT scans to show anatomical variations.

Post-op: Watch for bleeding. Post-op douches may be given ± topical steroids.

Nasal injury and foreign bodies

Fractured nose The upper $\frac{1}{3}$ of the nose has bony support; the lower $\frac{2}{3}$ and septum are cartilaginous. Direct violence to the nose may fracture the nasal bones. Take a history as to the time of the injury, previous nasal injuries, epistaxis, nasal obstruction, and csf rhinorrhoea. Ask about loss of consciousness. Consider other facial fractures: zygomaticomaxillary fractures may cause malocclusion of the teeth and impair normal jaw opening there may be diplopia if there is fracture of the orbital floor. Carefully feel around the orbit to check for 'steps' in contour. Radiographs are not particularly helpful as there may be old injuries shown and cartilaginous injuries do not show up. They will not alter the acute management.

Soft tissue swelling at the initial examination may make deformity difficult to see, so re-examine after 7 days (especially children)—exclude a septal haematoma at initial examination. If present, refer to an ENT surgeon for evacuation (risk of septal necrosis if untreated). Fracture reduction (under GA) is best within the first 10–14 days (complete setting may have occurred by 3 weeks). Zygoma and maxilla fractures set faster and should be referred immediately to maxillofacial surgeons. Post-operatively patients are advised to apply ice for the first 12h; sleep with head elevated, sneeze through mouth, refrain from blowing the nose and taking vigorous exercise. Re-examine at two weeks and two months after operation. Septal deviation may require septoplasty several months later. Post-traumatic nasal deformity is common (seen in 14–50%); this is reducible to 9% by complete nasal assessment (bony and septum), and primary septal reconstruction if severe septal fracture-dislocation.

medline:10946923

csf rhinorrhoea Fractures through the roof of the ethmoid labyrinth may result in leak of csf. Nasal discharge tests +ve for glucose (Clinistix® test, confirm with a laboratory sugar); csf uniquely contains β_2 -transferrin. Leaks often stop spontaneously; if not, try neurosurgical closure of the dura.

Septal haematoma This is rare but may be serious (beware rhinorrhoea; fever suggests abscess formation). These may occur after injury and cause distressing nasal obstruction, with bilateral septal swellings. If you detect a large, fluctuant boggy swelling in place of the septum, refer immediately. Evacuate clots at once under GA. Suture and pack as needed. Give co-amoxiclav (or clindamicin) to prevent infection. If left untreated there is risk of cartilage necrosis \pm nasal collapse.

medline:9557069

Foreign bodies Most are self-inserted by children. Organic material presents early with purulent discharge; inorganic bodies may remain inert for ages. If a child is co-operative it may be possible to grab the object with forceps. 2.5% cocaine spray may shrink the mucosa and allow extraction with a sucker. Foley catheters can also be useful. If general anaesthesia needed, protect the airway.

n = 69
medline:9002571

Cures of septal perforation After septal surgery (p 548); trauma; nose picking; inhalation of chrome salts; sniffing cocaine (look for other signs, below); rodent ulcer—or malignancy; or any chronic mucosal inflammation/granuloma—eg TB; syphilis; extra-GI Crohn's disease. Perforations irritate, crust, and bleed. *Treatment:* symptomatic. Closure is hard (may be done with rhinoplasty; a one-stage procedure).

medline:10935304

medline:10748848

Cocaine As well as septal perforation, inhaling (snorting) cocaine may cause odd behaviour, fever, and fits.

Diagnosis Blood levels are more time-consuming and expensive than qualitative methods (fluorescence immunoassay on urine). *

Nosebleed (epistaxis)

► *Never underestimate nosebleeds, they can be fatal.* Epistaxis is the commonest ENT emergency by far. It affects more than half the population at least once in their lifetime; while the vast majority will stop spontaneously or be easily controlled, in a minority it can be serious.

Causes In 80% the cause is unknown. The peaks in incidence occur in children, young adults, and those 50 or over. There is a seasonal variation: nosebleeds are more common in cold weather. 1064 medline:81351

Trauma (nose-picking; fractures) is a common cause; also ↓haemostasis eg blood dyscrasias, haemophilia, ↑alcohol intake. 50 medline:763452

Hypertension is not regarded as a cause, but it can prolong bleeding. Ask about NSAIDs & anticoagulant use (makes things harder). medline:850969

Conditions such as septal perforations and occasionally neoplasms can be the underlying cause. A search for the cause is often a secondary issue in the management; tests are only indicated for significant bleeds and routine clotting studies are not indicated. Don't waste too much time looking for rare causes (hepatic coagulopathy, Osler-Weber-Rendu syndrome, leukaemia, malaria, and typhoid etc). 140 medline:1078940

Management *First aid* Unless the patient is shocked, have them sitting up with the head tilted downwards to prevent blood trickling backwards. Firm pressure on the cartilaginous nose (not the bridge) using finger and thumb is needed for 10–15min.

- Protect yourself. It can be messy. Eye protection, gloves, and an apron.
- First, treat the patient. Assess for shock and resuscitate as needed.
- Then treat the nose. Find the bleeding and stop it. Direct visualization is the key to treatment but is often easier said than done. Bleeds are arbitrarily divided into anterior and posterior; anterior bleeds that can be easily seen with rhinoscopy are simpler to treat and are usually less severe. Always remember to check the oropharynx.

Anterior or 'simple' or minor', epistaxis This is almost invariably septal; Little's area is used to describe this area where ethmoidal, sphenopalatine, and facial arteries anastomose.

- 1 Remove clots with suction, or by asking him to blow his nose. Clot obscures the view, & contains fibrinolytic factors hindering haemostasis.
- 2 Insert a cotton wool pledget or a length of ribbon gauze soaked in a vasoconstrictor ± local anaesthetic, eg xylometazoline (Otrivine®) with 2% topical lidocaine—into the nostril and left for 5min. This will slow the bleeding and anaesthetize the area.
- 3 Obvious anterior bleeding can be cauterized with silver nitrate sticks. Warn that it stings. Apply firm pressure to, and around, the bleeding site for 10sec. Be careful with the clear fluid: it is corrosive.
- 4 Bleeding that persists can be treated with anterior packing with vaseline or BIPP (bismuth iodoform paraffin paste) impregnated ribbon gauze, or nasal tampons. Remember to attach a suture to the pack to secure it, and to aid removal.

If you cannot see the bleeding point, refer to an ENT unit. 15 medline:355339

Complicated epistaxis: (major or posterior epistaxis)

- Watch for shock especially in the elderly.
- Access is difficult and bleeding is usually more severe. Bleeding not localizable by anterior rhinoscopy, ie posterior epistaxis, can be treated by various methods depending on expertise and resources. The gold standard is visualizing the bleeding point eg with endoscopy, and then to directly treat, eg with bipolar cautery.
- If a bleeding site cannot be found, the nose may be packed with gauze. Bleeds arising more posteriorly require posterior packing: the simplest way is to pass a size 16G or 18G Foley catheter through the nose into the nasopharynx, inflate with 10–15mL water and then pull forward until it lodges to close off the posterior choanae. Place an anterior pack and remember to secure the catheter. Specialized balloon catheters such as Brighton balloons have an anterior and posterior balloon and are easier to use, but are less effective, as well as being more expensive. Inhalation of clot with respiratory obstruction has been reported with Brighton balloons. medline:8496648
- Packing is generally left for 48h; the patient is admitted and monitored. There are well-recognized problems with infections and hypoxia: infarcts, strokes, and deaths have been reported. Oxygen and pulse oximetry is recommended, and prophylactic antibiotics should be considered especially if packs remain for more than 48h—
toxic shock syndrome can occur (*OHCM* p592). medline:8233510

Persistent posterior epistaxis: Rebleeding after this can be treated by:

- **Examination under anaesthesia:** If a discrete bleeding point is found it can be treated directly, eg with diathermy otherwise repacking may be needed. Correction of septal deviation may improve access.
- **Ligation:** The emerging gold standard is endoscopic ligation of the maxillary/sphenopalatine artery around the sphenopalatine foramen. This is preferable to older techniques such as transantral arterial ligation of maxillary artery or ligation of the anterior ethmoidal artery. Embolization can be considered particularly for bleeding associated with tumours but can cause cerebrovascular accidents.

The paranasal sinuses

These are air-filled cavities in the bones around the nose; their exact function is unclear. They are lined by ciliated mucosa; debris and mucus are swept towards and through ostia into the nasal cavity.

Viruses cause mucosal oedema and depression of cilia action, leading to mucus retention which may become secondarily infected. Polyps and septal deviation can lead to poor drainage and so to sinusitis. Current emphasis in the understanding and treatment of recurrent/chronic sinus disease concentrates recognizing and correcting these drainage problems.

Sinusitis is generally divided into *acute* vs *chronic* types, distinguished primarily by their time characteristics but a true distinction is difficult. There are features common to both:

- Pain—eg maxillary (cheek; teeth)—or ethmoidal (between the eyes).
- Pain worse on bending.
- Discharge from nose, eg postnasal drip causing a foul taste in the mouth.
- Nasal obstruction/congestion
- Anosmia; ↓ smell; cacosmia (bad smell sensation without external source)
- Systemic symptoms, eg fever
- ▶ Swelling is uncommon: exclude carcinoma or dental root infections.

Differential diagnoses (non-sinus pain): migraine, TMJ dysfunction (p 535), neuralgias, cervical spine disease, temporal arteritis, herpes zoster.

Causes Most are to secondary bacterial infection following viral infection. 5% are secondary to dental root infection.

- Diving/swimming in infected water.
- Anatomical susceptibility (septal deviation, prominent uncinate process, large ethmoidal bulla, and polyps).
- General causes: Kartagener's, immunodeficiency, or general debilitation.

Images Combination of rigid endoscopy and CT (high definition) is the most sensitive. The latter generally being reserved for planning surgery, identifying the underlying problem but also to avoid the anatomical pitfalls eg low cribriform plates, vulnerable carotid arteries. Plain radiographs (occipitomental, occipitofrontal, and lateral views typically) and ultrasound are of little use. Normal radiographs cannot exclude sinus disease and 30% of normal sinuses show mucosal thickening. medline:9828797 & 8966269

Treatment *Acute sinusitis* Most are self limiting and need little further investigation. Bedrest, decongestants (short course), and analgesia may be tried. There is little evidence to support the use of antibiotics but these are often given (amoxicillin ± clavulanic acid). N 14
medline:9078199

If there is no response then drainage with lavage may be required.

Chronic recurrent sinusitis More likely to be related to structural/ drainage problems. Investigations should also consider general causes. The mainstay is *FESS* (functional endoscopic sinus surgery) tailored to the particular sinus(es) affected and the related anatomical problems. medline:10923936

Complications Much less common than previously, probably related to improved state of the nation's health ± antibiotic use. *Mucocoeles* (esp. frontal sinus) may become infected *pyocoeles*. *Orbital cellulitis/abscess* (this is an emergency, p 484). *Osteomyelitis* (classically staph, eg frontal bone).

Intracranial infection: meningitis, encephalitis, cerebral abscess, cavernous sinus thrombosis (fever, decreased conscious level and cerebral irritability).

The tonsils

Tonsillitis (sore throat \pm lymphadenopathy, especially jugulodigastric) often caused by Group A strep. Pathogens apart from streps: staphs, *Moraxella catarrhalis*, mycoplasma, chlamydia, haemophilus.

Differential diagnosis: Epstein–Barr virus (EBV), agranulocytosis, leukaemia, diphtheria, scarlet fever*.

Treatment: The use of antibiotics is a source of debate as in 50% the cause is viral, and antibiotics do not help. Typically penicillin v, 250mg/6h po. Despite studies advocating its use, amoxicillin is contraindicated: it causes a maculopapular rash in almost all whose pharyngitis is from EBV.

Indications for tonsillectomy

- Recurrent tonsillitis—exact definitions differ: a good general rule in children is >5 attacks lasting >5 days each year for the last 2 years. Controlled trials show that the incidence of sore throats after tonsillectomy is reduced, and a benefit from less time off school/ work.
- Airway obstruction, eg with sleep apnoea
- Chronic tonsillitis (>3 months, with halitosis)
- Suspicion of malignancy (unilateral enlargement/ ulceration)
- Quinsy

NB: some advocate 'hot' tonsillectomies ie during an acute attack particularly if association with a quinsy.

Local complications of tonsillitis. **Retropharyngeal abscess:** This is rare. Typical patient: an unwell child who fails to eat or drink. Lateral radiographs of the neck show soft tissue swelling. Treatment: Incision and drainage of pus under general anaesthesia in a head-down position to prevent aspiration.

Peritonsillar abscess (quinsy): This usually occurs in adults; odynophagia (painful swallowing) and inability to swallow saliva are cardinal symptoms. Trismus (lock-jaw) and swelling makes examination difficult. **Treatment:** incise/aspirate under local anaesthetic, or do abscess tonsillectomy under GA. Give antibiotics.

Complications The most significant complications of surgery is the rare but real risk of death from haemorrhage: primary (<24 hrs and usually requires a return to theatre) and secondary (typically 5–10 days and usually due to infection—here repeat surgery is rarely needed: it often settles with rest and antibiotics; friable tissues make haemostasis difficult and the pillars may need to be sewn together). Use of the tonsillar gag may cause damage to teeth, the temporomandibular joint, and posterior pharyngeal wall.

Differential diagnosis of unilateral tonsillar enlargement

- True asymmetry.
- Apparent enlargement—ie shift with peritonsillar abscess or parapharyngeal mass.
- Tumours: squamous cancers (70%), reticulum cell sarcoma, lymphosarcoma). Commonest in the elderly, presenting as sore throat, dysphagia and otalgia.

NB: unilateral tonsil enlargement is ominous: consider excision biopsy. Treatment: Radiotherapy, or surgery + cytotoxics or radiotherapy. In one study, survival for stage III & stage IV squamous cancers was 79 & 52%, respectively over 2–15yrs.

medline:10748454 & 8189757

***Scarlet fever:** The appearance of a rash after 48h on the neck and upper chest which spreads rapidly to abdomen and limbs suggests scarlet fever. It spares the mouth area, giving rise to contrasting circumoral pallor—and the tongue is covered with a white 'fur', which, when cleaned off, leaves prominent papillae (the 'strawberry' tongue). Cause: group A strep (treat with penicillin).

Sore throats*

In the past GPs readily gave antibiotics for sore throats in case the cause was a β -haemolytic streptococcal infection which may have resulted in rheumatic fever. Rheumatic fever is very rare in the West (p 194). We know that rheumatic fever patients had often had their sore throats treated, so antibiotics had not prevented the illness. GPs may not wish to give antibiotics for simple sore throats, as many are caused by viruses—in any case the proportion of those with sore throats consulting may be as low as 1 in 18. Because sore throat is so common, treating everyone with penicillin is not only expensive, and encouraging of doctor dependency (p 326), but also risks more deaths from anaphylaxis than would be saved by any possible benefit. Evidence that penicillin relieves strep throat symptoms is scant. With this in mind, throat swabs only have a limited rôle (NB: sensitivity 30%; specificity 80%—they have a rôle in teaching patients about their sore throats, and in allowing time to go by while nature effects a cure^{***}). Numerous antigen detection kits are available (specific, but not sensitive). When there is accompanying tonsillitis, there may be abdominal pain, fever, malaise, dysphagia, and painful neck lymph nodes.

A holistic approach to the person with a sore throat

Don't focus on the throat, the swab, or the microbiology: home in on people's health beliefs and work to harmonize these beliefs with your own.

Doctors often think patients expect antibiotics, and will be disappointed if they are not given. But frequently this is not the case. Does he attend with a very sore throat? If not, why now? 'My nephew is coming to stay, and my wife told me come and get a prescription'—or symptoms may be worse than usual. If so, ibuprofen may help. But what may *really* help is a dialogue between you and your patient. Strangely enough, there is evidence that such dialogue reduces symptoms rather than merely making them more acceptable.

www.rcgp.org.uk/rcgp/journal/issues/oct00/editor2.asp

But improving symptoms is not the only aim: your dialogue may promote patients' trust of their own body. It may be a stepping stone to active health rather than passive disease. It may stop your patient being a patient at all.

C Del Mar 2000 Cochrane Library Issue 3 & C Bradley 2000 *Br J Gen Pract* 50 781

The art of medicine consists of amusing the patient while nature cures the disease (Voltaire).

►► Stridor

Stridor is noisy inspiration due to partial obstruction at the larynx or distal large airways. Children develop airways obstruction more commonly and more dramatically than adults due to their airways being narrower and more readily deformed. (Adults tend to develop hoarseness).

Clinical features There may also be difficulty in swallowing, cyanosis, or pallor, use of accessory muscles of respiration, and downward plunging of the trachea with respiration (tracheal tug)—all of which are grave signs.

Causes *Congenital*: Laryngomalacia, web/stenosis, vascular rings. *Acquired*:

- **Inflammation**: laryngitis, epiglottitis, laryngotracheobronchitis, anaphylaxis.
- **Tumours**: haemangiomas or papillomas (usually disappear with onset of immunity—but may require laser treatment before).
- **Trauma**: thermal/chemical—or from intubation.
- **Miscellaneous**: airway or oesophageal foreign body; vocal cord paralysis.

Laryngotracheobronchitis (croup) is the commonest cause (much commoner than acute epiglottitis). 95% are viral—esp. parainfluenza (rib-avirin can help, eg if immunodeficiency: OTM 3e 338); bacteria include klebsiella & diphtheria; fungi are rare). Usually self-limiting, and treatable at home with humidification (steam) ± antibiotics. **Severity grading**: 1 Inspiratory stridor ± barking cough 2 Grade 1 + expiratory stridor 3 Grade 2 + pulsus paradoxus 4 Grade 3 + cyanosis or cognition↓. medline:11056758

Admit (eg to ITU) if severe. In children, CXR may show 'steeple sign' of a tapering trachea. Give antibiotics, humidified O₂, + nebulized epinephrine (5mL 1:1000, may buy time in severe disease needing ventilating), and prednisolone (may ↓ duration of intubation in adults, eg 1mg/kg/12h po).

Acute epiglottitis is a medical emergency as complete respiratory obstruction can occur. Typically the history is short, and the patient may become rapidly septicaemic. Admit to hospital: iv antibiotics, O₂ with a definitive airway. Don't examine the throat before admission as this may precipitate complete obstruction.

Adult epiglottitis is much rarer than croup—eg 9/10⁶/yr), but may have a higher mortality (mean 1.1%, but 17.6% if respiratory distress a presenting feature). Intubate early and benzylpenicillin + cefuroxime will cover the likely causes (*haemophilus*; *Strep pyogenes*). Get expert microbiological help.

Laryngomalacia (congenital laryngeal stridor) may appear within hours of birth. Floppy aryepiglottic folds and glottis ↑ tendency of the larynx to collapse in inspiration in affected infants. Stridor may be most noticeable in certain positions, during sleep, or when they are excited/ upset. In 85%, no treatment is needed and symptoms usually improve by 2 years but problems may occur with concurrent laryngeal infections. Parents need supporting with 3 months' follow-up. medline:10996231 & 10668673 & 10569405

Laryngeal paralysis A congenital paralysis accounting for 25% of infants with stridor—often the cause is unknown (but thought to be from vagal stretching at delivery), which may cause feeding difficulties. Their cry is usually normal as adduction is not affected. Most recover by 2–3 years.

Acute airway obstruction This may be overcome by skilled intubation, but if intubation fails, do needle cricothyrotomy in children (*OHCM* p 754)—jet O₂ at 15L/min through a wide-bore cannula (14G) placed in cricothyroid membrane: allow O₂ in for 1sec and exhalation (through partially obstructed upper airway) for 4sec. This technique can be used in adults. Alternative: surgical cricothyrotomy (not in children <12yrs)—see *OHCM* p 754. A tracheostomy in theatre is needed within 30mins as jet insufflation oxygenates rather than ventilates, so CO₂ builds up.

Hoarseness

Hoarseness is indicative of a vocal cord problem (structure and/or function). It is the commonest presentation of laryngeal carcinoma, and is often the only presenting complaint. Therefore, it is mandatory to investigate hoarseness (particularly in a smoker) lasting more than 3 weeks to exclude laryngeal carcinoma (p 562).

Other causes to consider Acute: Laryngitis; trauma (untrained singing, shouting, coughing, vomiting, inhaling noxious fumes); angioneurotic oedema and laryngeal abscess. *Chronic* (>3 weeks): Laryngitis; vocal cord paralysis; endocrine causes (acromegaly, Addison's disease, myxoedema); functional disorders; sicca syndrome (poor lubrication); granulomas, eg syphilis, TB (esp. if fever, cough, or weight loss), sarcoid, or Wegener's.

Investigations Laryngoscopy: assess cord mobility, inspect the mucosa, and exclude local causes.

Laryngitis This is usually viral and self-limiting, although there may be secondary infection with streps or staphs. NB: in *chronic* laryngitis, any bacteria found are more likely to be colonizers only. medline:11081617

Symptoms Pain (hypopharyngeal, dysphagia; pain on phonation), hoarseness; fever. Laryngoscopy reveals oedema.

Treatment Generally only supportive, but, if necessary, give phenoxymethylpenicillin 500mg/6h po for 1 week. Steam inhalations may also help. Chronic irritation of the cords from smoking or chronic voice misuse may lead to Reinke's oedema—a gelatinous fusiform enlargement of the cords (may also be associated with hypothyroidism)—if conservative measures fail, then the surgery usually took the form of vocal cord stripping, but now laser therapy is becoming popular.

Singer's nodules Caused by vocal abuse (untrained singers in smoky atmospheres). These are fibrous nodules classically at the junction of the anterior third and posterior two-thirds of the vocal cords. This is the middle of the membranous vocal folds (the posterior portion of the vocal fold is cartilage), and it may receive most contact-injury during speech. Nodules are often bilateral. **Treatment** Speech therapy may help in the early stages, or they can be removed surgically. www.txt.co.uk/full.cfm?1091...

Functional disorders Phonation produces no response in apparently normal vocal cord adductors—eg in young women at times of stress—but there are many functional disorders of the voice, which may result in laryngeal oedema ± nodules. The voice may be lost completely (aphonia) but is more commonly reduced to a whisper. A useful differentiating test is to ask the patient to cough (which requires functional adductors). The best treatment is speech therapy, with attention to the many undoubted emotional factors which may be present. NB: it is a diagnosis by exclusion—eg allergic reactions may cause sudden aphonia, so don't assume a functional disorder without laryngoscopy. medline:9747481...

Laryngeal abscess This is rare except in the immunocompromised (or post-trauma/intubation) and is difficult to diagnose (have a high index of suspicion, and remember TB). There is pain, fever, pain on swallowing (odynophagia), ± respiratory distress, and cervical lymphadenopathy. Lateral movement of the larynx is painful. Failure to respond to iv antibiotics (netilmicin with flucloxacillin to cover *pseudomonas*, staphs & *proteus*) or increasing respiratory distress warrants surgery. medline:10795598

Laryngeal nerve palsy

It is a common clinical finding that with partial paralysis cords are fixed in the midline, but in complete paralysis they are fixed mid-way (paramedian position). Proposed reasons for this include:

- Semon's Law: the nerve fibres supplying the adductors (posterior cricoarytenoid) are more susceptible to damage, and the midline position is due to unopposed adduction.
- Wagner Grossman theory: the cricothyroid (adductor) is supplied by the superior laryngeal nerve (external branch of) that arises more proximally and is thus more likely to be spared.

Causes: 30% are cancers (larynx in ~40%; thyroid, oesophagus, hypopharynx, bronchus, or any malignant node).

25% are iatrogenic, ie after (para-)thyroidectomy, oesophageal, or pharyngeal pouch surgery. Paralysis after thyroid surgery is seen in ≤5%, and is reversible in 35%.
medline:10911606

Other causes: CNS disease (polio; syringomyelia); TB; aortic aneurysm; 15% are idiopathic and are often ascribed to a neurotropic virus.

Tests: CXR, barium swallow/meal, MRI, panendoscopy.

Treatment (nonmalignant causes): Unilateral cord palsies can be compensated for by movement of the contralateral cord, but may require formal medialization in the form of fat/Teflon® injections or a thyroplasty, eg Isshiki-type.
medline:11012657

Tracheostomy may be needed to protect the airway.

ENT (head and neck) tumours

Nasopharyngeal cancer (beware unilateral ear effusion in adults) 25% of all malignancy in China vs 1% in UK. **Genetics** (abnormal HLA profiles) eg HLA A2 allele; there is ↓survival with B17 & BW46. **medline:2377207**

- Epstein-Barr (EBV) infection (there is increasing evidence). **medline:10781747**
- Weaning on to salted fish (?N-nitroso carcinogens). Its rôle may be overestimated—ingestion in quantity before age of 12 may be what is important.

Histology Squamous carcinomas (if keratinizing, risk factors are smoking and alcohol; if non-keratinizing, this is less of an association) and non-squamous: angiofibromas, lymphoepitheliomas, or lymphosarcomas.

Typical patient Usually early to upper deep cervical nodes. Local spread may involve cranial nerves via the foramen lacerum or jugular foramen. **Signs and symptoms** Epistaxis, diplopia, conductive deafness (Eustachian tube affected), referred pain, cranial nerve palsy (not I, VII, VIII), nasal obstruction, or neck lumps. **Diagnosis** Endoscopy & biopsy; PCR for EBV. **NB:** submucosal spread may mean the area looks normal. Stage by MRI, eg with STIR sequence (=short tau inversion recovery; better than CT). **medline:11077556**

Treatment Radiotherapy is mainstay ± chemotherapy ± surgery (radical neck dissection). 3-year survival for stage II is ~100% (93% for stage III; 69% for stage IV, in one study; worse if EBV +ve). **medline:11121629 & 11352952**

Acoustic neuroma These are slow growing, histologically benign lesions that cause problems by local pressure and then behave as a space-occupying lesion. It is a misnomer as it usually arises from the superior vestibular nerve schwann cell layer—hence the alternative name *vestibular schwannoma*. **The Patient** Progressive ipsilateral tinnitus ± sensorineural deafness, by compressing cochlear nerve. Big tumours may give ipsilateral cerebellar signs or ICPT signs. Giddiness is common; vertigo is rare. Trigeminal compression above the tumour may give a facial numbness. Nearby cranial nerves may be affected (esp. V, VI, VII). **Tests** MRI is best. The main differential diagnosis is meningioma. There may be a rôle for screening patients with unilateral tinnitus/deafness. **Treatment** Surgery is difficult; there are various approaches with different potentials for hearing preservation.

Pharyngeal carcinoma These are often advanced at presentation. ♂/♀≈5:1. Histologically, most (85%) are squamous. **Typical patient** Elderly smoker with sore throat, sensation of a lump, referred otalgia, and local irritation by hot or cold foods, with risk factors: chewing or smoking tobacco (alcohol alone is not a risk factor but is synergistic with smoking). 30% of squamous pharyngeal tumours will have a second primary within 10 years. 20% are lymph-node positive at presentation. Hypopharyngeal tumours may give dysphagia, voice alteration, otalgia, stridor, and throat pain; trismus is a late sign. Note any sign of premalignant conditions: leukoplakia and Patterson-Kelly-Brown syndrome (Plummer-Vinson)—in which a pharyngeal web is associated with iron deficiency, angular stomatitis, glossitis, and koilonychia: 2% risk postcricoid carcinoma. **Diagnosis** MRI with STIR (above) contrast-enhanced CT. **medline:10955293** **Treatment** Surgery, eg with endoscopic stapling ± later radiotherapy. Radiotherapy has a rôle as first-line therapy if the tumour is small, ie T1 (<2cm) and T2 (>2cm but <4cm). (T3 is >4cm and T4 beyond oropharynx.)

Sinus squamous cell cancer (~1% of all tumours) **Typical patient** Middle-aged or elderly. Suspect when chronic sinusitis presents for the first time in later life. **Signs and symptoms** Blood-stained nasal discharge and nasal obstruction. **Tests** Cheek swelling, swelling or ulcers of the buccal alveolar plate or palate, epiphora due to a blocked nasolacrimal duct, ptosis and

ptosis as the floor of the orbit is involved, and pain in maxillary division of the trigeminal nerve. Local spread may be to cheek, palate, nasal cavity, orbit, and pterygopalatine fossa. Patients present late because epistaxis, obstruction and headache only occur with large tumours. MRI/CT ± endoscopy (with biopsy) is best. NB: coronal CT is needed to show bone erosion (esp. around the cribriform plate). medline:10992946

Squamous cell (50%), lymphoma (10%), adenocarcinoma, adenoid cystic carcinoma, olfactory neuroblastoma, or chondrosarcoma, benign tumours.* Radiotherapy ± radical surgical. 5-year and 10-year overall survival rates are 77%. medline:11064349

Quantum cell laryngeal cancers 2000/yr in the UK.

Male smoker with progressive hoarseness, then stridor, difficulty swallowing ± haemoptysis or ear pain (if pharynx involved).

Regular cannabis users are also at increased risk. 2000 medline:11084527 & 3012440

Supraglottic, glottic, or subglottic. Glottic tumours have the best prognosis as they cause hoarseness earlier and spread to nodes is late.

Laryngoscopy with biopsy. MRI staging. medline:9393190

First-line options: Radical radiotherapy—or total laryngectomy ± neck dissection of neck glands. *If there is recurrence after radiotherapy:* Partial 'salvage' laryngectomy for some appears to be safe, effective, and results in reasonable preservation of laryngeal function. medline:10816221

Emotional costs: Salvage laryngectomy's economic and personal costs are great, especially in stage III & IV recurrences. Careful studies showing strong correlation of noneconomic costs with advanced stage allow creation of expectation profiles that may be useful to patients faced with difficult decisions. ▶ This needs to be a personal choice made 'after honest and compassionate discussion' with the surgeon. medline:10714711

Post-operative care: Patients have a permanent tracheostomy, so need to learn oesophageal speech. If a voice prosthesis is fitted at the time of surgery, reasonably normal speech is possible within weeks. Give pre-op counselling. Patients are usually discharged after ~10 days with a plastic vent or metal cannula to keep the tracheostomy open (discarded some weeks later). Excess secretions ± crusting around the stoma are common, needing meticulous attention—humidified stomal covers (eg Laryngofoam®) may get round these problems. Advise to take care while having a bath, and to avoid fishing and deep water (unless expert training to hand). Stenosis; recurrent pneumonia. Post-radiotherapy hypothyroidism (so monitor TSH). medline:10884190

Suggest a laryngectomy club. (UK: 0207 381 9993)

Hypopharyngeal tumours are rare and are usually a disease of the elderly. They can present as a lump in throat, dysphagia for solids then fluids, or as neck lumps. The anatomic limits of the hypopharynx are the hyoid bone to the lower edge of the cricoid cartilage: the 3 main sites are piriform fossa, postcricoid region, and the posterior pharyngeal wall. Males are more commonly affected except for postcricoid lesions. They are associated with smoking and alcohol, but not as clearly as laryngeal carcinoma. Treatment options are radiotherapy and surgery in various combinations. The prognosis is poor with 60% mortality at 1 year.

Inverted papillomas of the sinuses are the most important benign tumours of the sinuses. They are exophytic masses that show distinctive epithelial invagination of the stroma without destroying the basal lamina. Total excision can therefore be difficult without taking wide margins. Most significantly 2% show malignant change and 10% have a synchronous carcinoma.

Dysphagia

Dysphagia is difficulty in swallowing food or liquid. Unless it is associated with a transitory sore throat, it is a serious symptom: **further investigation is mandatory**, usually by endoscopy, to exclude neoplasia.

Painful swallowing is termed 'odysphagia'.

Globus pharyngeus is the sensation of a lump in the throat (even when not swallowing) with no obvious/physical cause and there is no true swallowing difficulty. The cause is unclear but reflux or motility disorder have been proposed: cricopharyngeal overactivity is sometimes demonstrated but whether this is primary or secondary is unknown. There is no specific treatment for this condition; reassurance is important. It is made worse by anxiety, and stress can form a vicious circle.

Causes Malignant:

Oesophageal cancer
Pharyngeal cancer
Gastric cancer
Extrinsic pressure
(eg lung cancer or
node enlargement)

Neurological causes:

Bulbar palsy (OHCM p 382)
Lateral medullary
syndrome
Myasthenia gravis
Syringomyelia (OHCM p 392)

Others:

Benign strictures
Pharyngeal pouch
Achalasia (below)
Systemic sclerosis
Oesophagitis
Iron-deficient anaemia

History and Examination. As there is usually little to find on examination, it is all in the history. There are 4 key questions to ask:

- 1 Can fluid be drunk as fast as usual, except if food is stuck?
Yes: Suspect a stricture (benign or malignant).
No: Think of motility disorders (achalasia, neurological causes).
- 2 Is it difficult to make the swallowing movement?
Yes: Suspect bulbar palsy, especially if he coughs on swallowing.
- 3 Is the dysphagia constant and painful?
Yes (either feature): Suspect a malignant stricture.
- 4 Does the neck bulge or gurgle on drinking?
Yes: Suspect a pharyngeal pouch (food may be regurgitated).

Tests: FBC; ESR; barium swallow; rigid endoscopy with biopsy; oesophageal motility studies (this requires swallowing a catheter containing a pressure transducer). CXR. Ambulatory pH studies have no proven value.

Nutrition Dysphagia can cause malnutrition. Nutritional support may be needed pre- and post-treatment, eg via a percutaneous endoscopic gastrostomy (PEG) tube. Get expert (dietitian's) help; see OHCM p 454. medline 1146423

Oesophageal carcinoma This is associated with achalasia, Barrett's oesophagus (OHCM p 720), tylosis (a hereditary condition in which there is hyperkeratosis of the palms), Patterson-Brown-Kelly (or Plummer-Vinson syndrome, p 562) and smoking. Survival after resection is rare after 5 years (OHCM p 492).

Benign oesophageal stricture Causes: oesophageal reflux; swallowing corrosives; foreign body; trauma. **Treatment:** Dilatation (endoscopic or with bougies under anaesthesia).

Barrett's oesophagus See OHCM p 720.

Achalasia There is failure of oesophageal peristalsis and failure of relaxation of the lower oesophageal sphincter. Liquids and solids are swallowed slowly. CXR: air/fluid level behind the heart, and double right heart border produced by an expanded oesophagus. Typical 'rat's tail' appearance. **Treatment:** Myomectomy cures ~75%. Pneumatic dilatation may also help.

Facial palsy

Arising in the medulla, and emerging between pons and medulla, the facial nerve passes through the posterior fossa and runs through the middle-ear before emerging from the stylomastoid foramen to pass into the parotid. Lesions may be at any part of its course. Branches in the temporal bone:

- 1 The greater superficial petrosal nerve (lacrimation),
- 2 Branch to stapedius (lesions above this cause hyperacusis),
- 3 The chorda tympani (supply taste to anterior $\frac{2}{3}$ of the tongue).

Causation: Brainstem tumours; strokes; polio; multiple sclerosis; cerebellopontine angle lesions (acoustic neuroma, meningitis).

Causes: Otitis media; Ramsay Hunt syndrome; cholesteatoma.

Infratemporal: Parotid tumours; trauma.

Causes: Lyme disease; sarcoid; Guillain-Barré; herpes; diabetes; Bell's palsy.

Examination and tests Lower motor neurone lesions can cause paralysis of the whole of one side of the face, whereas in upper motor neurone lesions, the facial muscles of the forehead may still be operational (they are bilaterally innervated). Brainstem lesions produce only muscle weakness and may be accompanied by sixth nerve palsies. Loss of lacrimation (Schirmer's test, p 482), stapedius reflex, taste (electrogustometry detects) and ↓sub-mandibular saliva production (cannulate ducts) imply nerve lesions proximal to the origin of the relevant branches. Examine the ears (to exclude otitis media, zoster, and cholesteatoma) and parotid. Consider temporal bone radiography. Electromyography reveals completeness of the lesion.

Management Examine the facial nerve in victims of head trauma. An incomplete palsy will probably recover. A complete palsy demands surgical exploration.

Infection In acute otitis media accompanied by facial nerve palsy myringotomy should be performed. When cholesteatoma is present emergency exploratory surgery should be performed. If rare acute necrotizing otitis externa is the cause, iv antibiotics (eg ticarcillin) and local toilet are needed.

Ramsay Hunt syndrome (Herpes zoster oticus) See p 756.

Bell's palsy At present the cause is unknown but many believe it part of a viral polyneuropathy (typically Herpes simplex in some small studies) with demyelination, the V, X, and C2 nerves also being affected. Onset is abrupt and may be associated with pain. The mouth sags, and dribbling, taste impairment and watering (or dry) eyes may occur. The patient cannot wrinkle the forehead, blow forcefully, whistle, or pout out his cheek.

Management Protect the eye with dark glasses, and tape closed during sleep. If recovery is expected to take months, consider lateral tarsorrhaphy. Instil artificial tears regularly at the slightest evidence of drying. There is good evidence that prednisolone helps, *if given early*¹ (eg in first 24h, eg 20mg/6h for 5 days, tail off over next 5 days). Oral aciclovir (eg 400mg five times a day) appears to be a promising *additional* treatment.² Hooks and cheek plumpers may improve appearance. If electromyography reveals total degeneration, surgical exploration may be carried out to check nerve continuity, breaches of which may be grafted (eg with lateral cutaneous nerve of the thigh).

1 Many neurologists give steroids 'to reduce oedema' particularly to those seen within 6 days of onset. One helpful study is that by TS Shafshak (1994 *J Laryng & Otology* 108 940-3 & Bandolier 1995 2/11 3), showing that the extra benefit of steroids may be confined to those treated within 24h of onset. Spontaneous recovery is good in any case (85%). For every 3 persons treated with steroids within 24h 1 extra had a good recovery compared with no treatment; for ethical reasons, this study was not randomized. Older randomized studies have been inconclusive, but did not look specifically at early treatment. A recent meta-analysis supports the use of steroids (I Williamson 1996 *Br J Gen Pract* 743-7 8 E-8M 1997 2 79.) 2 R Enting 1998 *Ned Tijdschr Geneeskde* 142 436

Lumps in the neck

► **Do not biopsy a neck lump.** Any possibly malignant neck lump should be referred to ENT, where thorough assessment and search for a primary may be undertaken, possibly in a dedicated 'neck lump' clinic, with access to fine-needle aspiration (FNA) for cytology (beware pulsatile lumps!), and CT/MRI (MRI is better). Remember to culture any specimen for TB.

Diagnosis • **How long has the lump been present?** If <3 weeks, lymphadenopathy from a self-limiting infection is likely, and extensive investigation is unwise. • **Which tissue layer is the lump?** Is it intradermal? (eg from sebaceous cyst with a central punctum, or a lipoma).

If the lump is not intradermal, and is not of recent onset, allow yourself to feel the intoxicating sensation of a hunt starting off over complicated terrain.

Midline lumps If patient <20yrs old, the likely diagnosis is a **dermoid cyst**, or, if it moves on protruding the tongue and is below the hyoid, a **thyroglossal cyst** (fluctuant lump developing in cell rests in thyroid's migration path; treatment: surgery). If over 20, it's probably a thyroid mass, unless it's bony hard, when the diagnosis may be a chondroma. medline:10086242

Submandibular triangle (Below jaw; above anterior belly of digastric.) If <20yrs, self-limiting lymphadenopathy is likely. If >20, exclude **malignant lymphadenopathy** (eg firm, and non-tender). ► Is TB likely? If it's not a node, think of submandibular **salivary stone, tumour, or sialadenitis**. medline:11002367

Anterior triangle (Below digastric and in front of sternomastoid.) Nodes are common (see above): remember to examine the areas which they drain (Is the spleen enlarged?—this may indicate lymphoma). **Branchial cysts** emerge under the anterior border of sternomastoid where the upper 1/3 meets the middle 1/3; age <20. The popular theory is that they are due to non-disappearance of the cervical sinus (where the 2nd branchial arch grows down over 3rd and 4th) but this is not universally accepted. Lined by squamous epithelium, their fluid contains cholesterol crystals. Treat by excision. **Cystic hygromas** arise from the jugular lymph sac and transilluminate brightly. Treat by surgery or hypertonic saline sclerosant. **Carotid body tumours** (chemodectoma) are very rare, move from side-to-side, but not up and down, and splay out the carotid bifurcation. They are firm (softness is rare) and pulsatile, and do not usually cause bruits. They may be bilateral, familial, and malignant (5%). This tumour should be suspected in tumours just anterior to the upper third of sternomastoid. Diagnose by digital computer angiography. Treatment: extirpation by vascular surgeon. If the lump is in the superoposterior area of the anterior triangle, is it a **parathyroid tumour**? (more likely if >40yrs). A **laryngocele** is an uncommon cause of a lump in the anterior triangle: it is painless, more common in males, and is made worse by blowing. medline:3180497

Posterior triangle (Behind sternomastoid, in front of trapezius, and above the clavicle.) If there are many small lumps, think of **nodes**—TB or viruses, eg HIV or EBV (infectious mononucleosis) or, if over 20yrs, consider lymphoma or metastases. The primary may be head and neck (eg tongue base, posterior nasal space, tonsils, etc) or bronchus, gut, breast, or gonad (in that order of likelihood). **Cervical ribs** may intrude here. medline:11115294

Tests Ultrasound shows lump consistency. CT defines masses in relation to their anatomical neighbours. Do virology and Mantoux test. CXR may show malignancy or reveal bilateral hilar lymphadenopathy, when you should consider sarcoid. Consider fine-needle aspiration (FNA).

The salivary glands

► Refer all patients with salivary gland swelling.

The 3 major pairs of salivary glands are: *parotid, submandibular & sublingual*.

History and examination Dry mouth/eyes; lumps; swelling related to food; pain. Look for external swellings, secretions (in mouth first), do bimanual palpation for stones, test VII nerve, any regional nodes?

Inflammation is usually due to infection or an obstructing calculus (calcium phosphates and carbonates form around a nidus of cells and organisms).

Classification Are symptoms unilateral, or bilateral? Acute or chronic?

Acute unilateral symptoms are usually due to mumps (p 214). The patient is usually young and complains of parotid swelling.

Acute, unilateral symptoms are also likely to be due to mumps but acute parotitis may occur by ascending oral infection. This occurs post-op but is now rare, unless dehydrated or poor oral hygiene. **Treatment** Oral hygiene, hydration, antibiotics (depends on culture and sensitivities). If the duct stops draining pus, or pain and pyrexia increase, there may be abscess formation which will need draining through the skin.

Recurrent unilateral symptoms are likely to be due to stones (submandibular in 80%). The classic story is of pain and swelling on eating. The gland may be red, swollen, and tender (not necessarily infected). Stones may be seen on plain radiography or by sialography. **Treatment**. Distal stones are removed via the mouth but excise the gland if it contains the stone.

Chronic bilateral symptoms may be associated with dry eyes and mouth ± Sjögren's or Mikulicz's syndromes (OHCM p 730–4). **Treatment** is hard. If chronic infection is the cause, simple antibiotics will fail: what is needed is prolonged treatment with oxytetracycline 250mg/12h 1h ac po.

Fixed swellings may be idiopathic or due to sarcoidosis.

Tumours '80% are in the parotid, 80% of these are pleomorphic adenomas, 80% of these are in the superficial lobe.' * Any salivary gland swelling must be removed for assessment if present for >1 month (or examined by fine needle aspiration for cytology: this does not lead to seeding along its track). VII nerve palsy suggests malignancy. As tumours grow by budding and have no capsule, lumpectomy leads to seedling deposits, so superficial parotidectomy is needed. NB: sialograms with CT (+bone filters) may be a useful pre-op test. Classification is complex: full details in the electronic version. ⁶²

Classification Benign:	Intermediate:	Malignant:
Pleomorphic (mixed parotid) adenoma	Mucoepidermoid tumours	Adenoid cystic carcinoma
Adenolymphoma (Warthin's tumour)	Acinic cell carcinoma	Adenocarcinoma
Haemangioma/lymphangioma (child)	Oncocytoma	Squamous cell carcinoma

Middle age; slow growth; must be removed by superficial parotidectomy or enucleation. Radiotherapy has a rôle if there was intraoperative spillage, or in residual disease, or recurrences (seen in ~1–2% at 12 years follow-up). Post-op complications: metastatic spread (rare); facial nerve palsy (with skill, risk is minimal); gustatory sweating (~7%).

Usually elderly men; soft. Enucleation.

Rapid growth; hard fixed mass; pain & VIIth nerve palsy. Treat by combination of surgery and radiotherapy.

VIIth nerve palsy—often transient.

Salivary fistula—often closes spontaneously.

(gustatory sweating); here tympanic neurectomy may help

Dry mouth (xerostomia)

Sources: www.doctorupdate.net/du_reference/du_refarticle.asp?ID=6209#

- Dry, atrophic, fissured oral mucosa
 - Discomfort, leading to difficulty eating, speaking, and wearing dentures.
 - No saliva pooling in floor of mouth
 - Difficulty in expressing saliva from the major ducts
- When examining for the cause, check for salivary gland swelling.

Complications:

- Dental caries
- Candida infection

Causes

- Drugs: many, eg tricyclics, antipsychotics, β -blockers, diuretics, hypnotics
- Mouth breathing
- Dehydration
- Head and neck radiotherapy
- Sjögren's syndrome, SLE, scleroderma
- Sarcoidosis
- HIV/AIDS
- Obstruction (eg parotid sialolith; only 40% are radio-opaque vs >80% in submandibular glands)
- Graft-versus-host disease

Management:

- Increase oral fluids; take frequent sips.
 - Good dental hygiene; avoid acidic drinks/foods that demineralize teeth.
 - Try a saliva substitute, eg sst[®] tablets (contains polyethylene and cottonseed oil). 'Allow a tablet to dissolve in the mouth—while moving it around with your tongue.' Up to 16 tablets per day may be needed.
 - Chewing sugar-free gum or sweets may ↑ salivary flow. medline:11404944
 - Pilocarpine (Salagen[®]) is rarely satisfactory. medline:11336078 & 11819900
- Cholinergic SEs: sweating, lacrimation, rhinitis, amblyopia, diarrhoea, urinary frequency. Pilocarpine mouth washes may obviate these. medline:11743622
- Irradiation xerostomia: Acupuncture may help, eg as an 8-needle regimen of 3 weekly sessions followed by monthly sessions. medline:11920486

Dentistry for doctors

Any oral ulcer which has not healed in 3 weeks should receive specialist assessment, for biopsy to exclude malignancy (OHCM p 216).

Causes of facial pain: Tooth pathology, sinusitis, temporo-mandibular joint (TMJ) dysfunction, salivary gland, migraine, trigeminal neuralgia, atypical facial pain (no obvious organic cause), trauma, cluster headaches (OHCM p 331) myocardial infarction, frontal bone osteomyelitis (post sinusitis), ENT tumours

When helping a patient with a dental infection pay attention to these features—before consulting a maxillofacial surgeon, or a dentist (GDP).

1 *Is it the teeth?* History: *Is the pain...*

- Worse with sugar and heat?
 - Worse or better with cold?
 - Intermittent?
- } ▶ Tooth is alive (pulpitis)

Is the pain...

- Worse with percussion?
 - Constant/uninterrupted?
- } ▶ Tooth dead (osteitis/abscess)

Is the pain...

- Exacerbated by movement } ▶ Abscess between finger and thumb (>1mm)

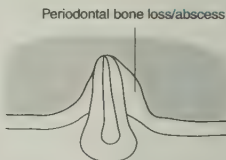
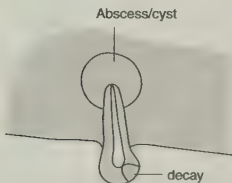
Radiography (usually helpful): Orthopantomogram (OPT) is useful for imaging molars and pre-molars. If incisors are suspected, request periapical radiographs of the tooth in question. Interpretation of radiograph...

• Abscess

(Tooth tender to percussion)

• Periodontal disease

(Tooth mobile)



(Opening mouth is difficult because of spasm or pain.) This is a sign of severe infection. Ask the patient to open mouth wide and measure how many fingers breadth between the incisor teeth. Trismus always requires maxillofacial advice. Other causes: tetanus; neoplasia.

Usually subside with oral antibiotics. If swelling is related to the lower jaw, assess for airways obstruction; if spreading to the eye, assess the second cranial nerve. If in any doubt, refer to a maxillofacial surgeon.

Temperature (very important), pulse and blood pressure. This information must be to hand prior to referral. Systemically unwell patients require maxillofacial advice/admission.

Any immunocompromise (eg HIV, leukaemia, diabetics, those on steroids); patients at risk of endocarditis; coagulopathy (eg haemophilia or warfarin). Seek specialist advice. In one HIV study, lesions included: candidiasis (29%), ulcers (15%), salivary gland disease (9%), necrotizing ulcerative gingivitis/periodontitis (5%), linear gingival erythema (4%), labial molluscum contagiosum (3%), oral warts (2%), hairy leukoplakia (2%), and herpes zoster (1%).

Dental caries Although on the decline in the West due primarily to fluoride, this condition is increasing exponentially in developing countries. Causes: bacteria (esp. *S mutans*), substrate (sugars) and susceptible tooth surface. In otherwise healthy individuals it is an entirely preventable disease.

Rampant caries is a variant found in children exposed to excessive sucrose in the forms of coated dummies, 'health' drinks, and prescribed medicines.

Radiation caries (eg post head & neck radiotherapy, eg with jaw osteoradionecrosis). *Treatment and complications:* Pain \pm infection. Toothache pain responds best to NSAIDs, eg ibuprofen 200–400mg/8h po pc (can buy it from pharmacists), and dental infection to penicillin and metronidazole, but drug treatment is never definitive, and a dental referral is required.

Periodontal disease 'Is one of the most widespread diseases of mankind' (WHO). Virtually the entire dentate adult population has gingivitis which is caused by bacterial and polysaccharide complexes at the tooth-gingival interface (=plaque). It is cured by good toothbrushing. Acute herpetic and rarely streptococcal infection can cause gingival inflammation.

Vincent's angina is a smoking or HIV associated painful, foul-smelling, ulcerative gingivitis, caused by anaerobes (*Fusobacteria*) \pm spirochaetes (*Borellia vincentii*). Give penicillin 250mg/6h po + metronidazole 200mg/8h po.

Gingival swelling Fibrous hyperplasia may be congenital or via drugs (phenytoin, ciclosporin, nifedipine). Other causes: pregnancy, exacerbation of inflammation by immunodeficiency, scurvy, infiltration in leukaemia.

Periodontitis (pyorrhea) is a progression of localized inflammation from the gums into the ligament supporting a tooth. It is associated with anaerobic bacteria, calcified bacterial deposits (calculi, tartar) and poor oral hygiene. It requires treatment by a dentist.

Aggressive periodontitis may be caused by lack of nutrients, an immune problem, neutropenia, or an intrinsic neutrophil disease—leukocyte adhesion deficiency, Chediak–Higashi or Papillon–Lefevre syndromes (with palmar keratosis), chronic granulomatous disease. medline:11692824 & 11452908 & 11189956

Malocclusion Inappropriate positioning of the teeth in the jaws or between the jaws themselves is common. Those with prominent upper teeth are particularly prone to trauma, and those children at risk (eg in epilepsy)—or those involved in contact sports should be referred to an orthodontist. Those with severe facial or jaw disharmony who may be unable to chew or have psychological difficulty with their appearance may be amenable to surgical correction by an oral or maxillofacial surgeon.

Wisdom teeth In the West, these can be considered, rather like the appendix, as a vestigial organ. They account for an enormous number of outpatient operations annually. If asymptomatic and not exposed to contamination by the mouth, they do not usually require removal. Complications are pain and infection and they may be involved in fractures of the mandible. Post-operative recovery is often complicated by pain and swelling, pain responds well to NSAIDs and poorly to opiates, particularly hydrocodone. Infection complicates up to 30% not receiving antibiotics. Penicillin and metronidazole (as above) are standbys.

Teething An acute sore mouth during tooth eruption is often caused by viral infections (eg herpetic). The onset of eruption of first deciduous teeth correlates with the fall off in transferred maternal antibody.

NB: Other oral mucosal diseases are described in *OHCM* p 216

8. Dermatology

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Acknowledgement: We particularly thank Dr Susannah Baron for masterminding changes to this chapter, and for supplying images.

Downloaded from www.freebooks4doctors.com; images compendium: www.dermis.net/doia/mainmenu.asp?zugr=d&lang=e

MacKie Rona M. *Clinical Dermatology*, 4e 1997, OUP, ISBN 0-19-2627619, and Rook, Wilkinson/Ebling *Textbook of Dermatology* 5e OUP, ISBN 0-632-02396-1. www.cochrane.org see the Cochrane Skin Group.

medline:9875203 & 10791153

An introduction to dermatology

The skin is one of the largest organs in the body and can display a myriad of clinical signs and reaction patterns. Recognition of these by the clinician may permit diagnosis of previously unsuspected systemic disorders. Primarily dermatological conditions such as eczema and psoriasis are not only the domain of dermatologists but are likely to be encountered by all clinicians, regardless of field of practice. A practical knowledge and clinical confidence in the diagnosis of skin disease is thus a most valuable asset.

Dermatologists do not confine themselves to the skin. Skin symptoms are features of many medical (p 580) and psychiatric illness, eg body image problems, dermatitis artefacta, neurotic excoriations, and trichotillomania (neurotic pulling out of one's hair). medline:11843209

Psychiatric aspects of skin disease There is an association between major depression/social phobia and psoriasis, and between obsessive compulsive disorder/stress and acne. Anyone who doubts the metaphysical relevance of psychocutaneous phenomena need look no further than Dennis Potter's *Singing Detective*, Philip Marlow.*

In acne, the psycho-physiology is interesting. Sebaceous glands and their secretions may be involved in a pathway conceptually similar to the hypothalamic-pituitary-adrenal axis, so mediating a direct link between oily skin, and the stress hormone CRH (corticotrophin releasing hormone) which acts on sebaceous glands. So think of CRH as an autocrine hormone for human sebocytes that exerts homeostatic lipogenic activity, with testosterone and growth hormone inducing CRH negative feedback. These findings implicate CRH and, therefore, stress in the development of acne, seborrhoea, androgenic alopecia, skin ageing, and xerosis. Substance P is another route connecting stress, seborrhoea, and acne. medline:12011471 & 11479771

Telemedicine, virtual care, and the Internet

Traditionally the dermatologist sees patients on wards and in central clinics: convenient for him or her, but less convenient for patients who live far away, and this paradigm does not allow for useful interaction between the referring doctor and the specialist. Also, urgency cannot be assessed until the patient presents (unless a photograph is sent with the referral letter; this simple idea in fact requires e-mail, digital cameras, and technical commitment). Telemedicine uses high-quality video links and computer equipment to form a virtual consultation. Is the expense entailed worth it? What are the problems?

Telemedicine in dermatology medline:12020415 & Lancet 1957 & 1961

Travel times are less	Expense is not outweighed by ↓travel time
Fewer tests ordered	Consultant does not share in patients' problems
Teaching referring GPs	Technical problems can waste whole sessions
Patient satisfaction	Extra time needed from GPs and consultants
Fewer follow-ups	Important clinical cues are lost in telemedicine

Medline:9279935 & <http://bmj.com/cgi/content/full/315/7123/1709>

History and examination

History Duration of rash; site of onset, spread, and distribution of lesions; symptoms (itch, pain); aggravating factors (sunlight, heat); previous treatments; medical conditions and medications; family history (psoriasis, atopy); occupation (industrial chemicals); pets.

Examination Examine all the skin; hair and nails where appropriate.

Distribution Symmetrical flexural (atopic eczema—PLATE 8.8); contact with jewellery or cosmetics (allergic contact dermatitis); areas exposed to sun, eg backs of hands, face, neck (photosensitivity); grouped lesions (herpes virus); symmetrical extensor surfaces (psoriasis).

Pattern Ring (fungal—active edge with healing centre, p 590); linear (Köbner phenomenon, below), targetoid (erythema multiforme).

Terms used to describe lesions www.pediatrics.wisc.edu/derm/tutorials.htm

alopecia hair loss

atrophy thinning and loss of skin substance

bulla blister larger than a vesicle (see below, ie >0.5cm diameter)

crust dried brownish exudate

erosion superficial break in the continuity of the epidermis; healed without scarring

erythema reddening of the skin which blanches on pressure

excoriation scratch which has broken the surface of the skin

hyperkeratosis long, irregular projections, which may be threadlike, or broader, like close-packed tombstones on a mound (seen in warts)

fissure crack, often through keratin

induration an area of thickening of the dermis and subcutaneous tissues

injury skin lesions which develop at a site of injury (psoriasis)

lichen lichen planus, plane warts, vitiligo www.txt.co.uk/full.cfm?111

leukoderma skin thickening with exaggerated skin markings, as a result of repeated rubbing (PLATE 8.10)

macule flat well-defined area of altered skin pigmentation; big lesions are called **patches** (*macula peccati* means the stain of original sin)

nodules solid, mostly subcutaneous lesion, more than 0.5cm diameter

papule raised well-defined lesion, usually less than 0.5cm diameter

plaque raised flat-topped lesion, usually over 2cm in diameter

purpura purplish lesion resulting from free red blood cells in the skin. It does not blanch on pressure

pustule well-defined pus-filled lesion

scale fragment of dry skin

scar permanent replacement of skin area with connective tissue

scarification loss of epidermis and dermis which results in a scar

vesicle blister less than 0.5cm diameter

wheel transient raised lesion with a pale centre and pink margin

Creams, ointments, and how to use them

The word 'topical', like 'topography', pertains to *surfaces* (from the Greek, *topicos*), so when you are told by a dermatologist to think of a topical treatment, he or she is not referring to current affairs but to treatments which are applied directly to the skin. In general, ointments are for dry areas, and creams are for moist areas. Ointments are greasy, and contain no added water, whereas creams are oil-in-water or water-in-oil emulsions. (Lotions are water-based and may be made up from powders: they are less commonly used, but may be tried as a coolant—calamine lotion, for example.)

If a large area of skin is involved, a large volume of cream or ointment will be needed, eg 500g.

Use simple clean procedures when getting the cream out of the pot, eg removing it with a clean spoon first. Tubs of ointment should be closed after use. Pump dispensers are a good way around bacterial contamination.

In dermatology, regimens may be complex, and you may get annoyed at the patient's 'poor compliance'—until you reframe this as loss of *concordance*. Concordance means there is harmony between your view of what should be done, and the patient. So plan treatments through dialogue, understanding the constraints his or her lifestyle may impose. The dermatology or practice nurse may be invaluable in demonstrating how to use topical therapies, and in optimizing concordance.

Possible causes of patterns of skin disease

Ring-shaped (annular) lesions

www.geocities.com/NapaValley/4122/derm.htm

- Fungal infections: active erythematous scaly edge with central healing.
- Erythema multiforme: target-like lesions, eg on extensor limb surfaces
- Granuloma annulare: pinkish papules forming a ring.
- Basal cell carcinoma (rodent ulcer; PLATE 8.5): pearly papule + central ulcer

Round, oval, or coin-shaped (discoid) lesions

- Psoriasis: red-pink plaques covered with a silvery scale (PLATE 8.11-B).
- Discoid eczema: well-defined crusted eczematous lesions.
- Pityriasis rosea: herald patch; oval red lesions with scaly edge, eg on trunk
- Impetigo: well-defined red patches, covered with honey-coloured crust.

Linear lesions • Köbner phenomenon: psoriasis, lichen planus.

- Dermatitis artefacta: linear or bizarre-shaped lesions, induced by patient.
- Herpes zoster: polymorphous vesicles/pustules in dermatomal distribution.

White lesions • Pityriasis versicolor: superficial slightly scaly infection

- with the yeast *Malassezia furfur*; appears depigmented on darker skins.
- Pityriasis alba: post-eczema hypopigmentation, often on children's faces.
- Vitiligo (*vitellus* is Latin for *spotted calf*: typically white patches ± hyperpigmented borders. Sunlight makes them itch. *Associations*: Autoimmunity; pernicious anaemia, thyroid or Addison's disease—also diabetes, alopecia areata, hypoparathyroidism. *Treatment* (unsatisfactory): cosmetic camouflage may help. PUVA (p 587) or potent topical steroids are sometimes helpful.
- Rarely, leprosy.

www.txt.co.uk/full.cfm?1115

Brown pigmented lesions • Freckles: brown macules, related to sun exposure.

- Lentigos: persistent brown macules, often larger than freckles.
- Café-au-lait spots: faint brown macules; if >5, consider neurofibromatosis.
- Seborrhoeic keratoses/warts (PLATE 8.7): benign greasy-brown warty lesions usually on the back, chest, and face; very common in the elderly.
- Chloasma (melasma) are brown patches especially on the face, related to pregnancy or Pill use. The condition may respond to topical azelaic acid.
- Systemic diseases: Addison's (palmar creases, oral mucosa, scars); haemochromatosis; porphyria cutanea tarda (+ skin fragility and blisters).

Subcutaneous nodules Rheumatoid nodules, rheumatic fever, PAN, xanthelasma, tuberous sclerosis, neurofibroma, sarcoid, granuloma annulare.

Itch (pruritus) • Itch can be very distressing. Skin will usually be scratched or rubbed and a number of secondary skin signs are seen: excoriations (scratch marks); lichenification (skin thickening, PLATE 8.10); papules or nodules (localized skin thickening). (Dry skin tends to itch.) Determine if there is a primary skin disease or is itch due to a systemic disorder.

- Itchy skin disorders: scabies (burrows in finger-webs, wrists, groins, buttocks); urticaria (transient weals, dermatographism); atopic eczema (flexural eruption, lichenification—PLATE 8.8); dermatitis herpetiformis (very itchy blisters on elbows, shoulders); lichen planus (flat topped violet papules, especially on wrists). www.txt.co.uk/full.cfm?1112
- Medical conditions causing itch: iron deficiency (koilonychia, pale conjunctivae); lymphoma (lymphadenopathy, hepatosplenomegaly); hypo/hyperthyroidism (goitre); chronic liver disease (jaundice, spider naevi); chronic renal failure (dry sallow skin); malignancy (clubbing, masses); adverse drug reactions. Investigations of itch in the absence of skin disease should include: FBC, ESR, ferritin, LFT, U&E, glucose, T₄, CXR.

Treat any primary disease; bland emollients (eg Diprobase®) to soothe dry skin; emollient bath oils; sedative antihistamines at night.

Skin manifestations of systemic disease

Diabetes mellitus Flexural candidiasis; necrobiosis lipoidica (waxy, shiny brownish plaques on shins); folliculitis; skin infections. www.bxt.co.uk/full.cfm?111

Celiac disease Dermatitis herpetiformis (very itchy/burning blisters on the elbows, scalp, shoulders, and ankles. Responds quickly to dapsone (50–200mg/day po). SE: haemolysis, LFT↑, agranulocytosis. CI: G6PD↓. Gluten-free diet may clear lesions. Small risk of lymphoma with chronic disease.

Inflammatory bowel disease • Erythema nodosum (tender ill-defined subcutaneous nodules, usually on the shins. Other associations: sarcoid drugs, TB, streptococci) • Pyoderma gangrenosum: recurring nodulo-pustular ulcers, ~10cm wide, with tender red/blue overhanging necrotic edge, healing with cribriform (pitted) scars. Site: eg leg; abdomen; face. Other associations: autoimmune hepatitis; neoplasia; Wegener's; myeloma. ♀:♂ >1:1. Differential: Behçet's, warfarin necrosis, rheumatoid vasculitis, 3° syphilis, blastomycosis, skin amoebiasis. Treatment: Get expert help. Saline cleansing high-dose oral or intralesional steroids ± ciclosporin ± topical antibiotics.

Rheumatoid arthritis Rheumatoid nodules; vasculitis (nodular, palpable purpura, with ulceration); pyoderma gangrenosum. www.bxt.co.uk/full.cfm?111

Systemic lupus erythematosus Facial butterfly rash; photosensitivity (face, dorsum of hands, V of neck); red scaly rashes; diffuse alopecia. Diagnosis: circulating autoantibodies; skin biopsy with immunofluorescence.

Thyroid disorders Pre-tibial myxoedema (diffuse thickened pink nodules/plaques on shins); clubbing (thyroid acropachy); diffuse alopecia. Myxoedema: Sparse coarse hair; dry skin; eczema craquelé (emollient-responsive crazy-paving, asteatotic eczema, also seen on old shins). www.bxt.co.uk/full.cfm?1106

Neoplasia Velvety, pigmented, thickened skin, eg in axillae; associated with adenocarcinoma of stomach. www.bxt.co.uk/full.cfm?1103

Dermatomyositis Purple rash, eg on eyelids/face, associated with muscle weakness. Exclude lung, breast, and ♀ GU tract cancer. www.bxt.co.uk/full.cfm?1104

Sjögren's disease Generalized dry skin; recent onset can point to underlying lymphoma or carcinoma. See p 578. www.bxt.co.uk/full.cfm?1105

Erythema multiforme Minor form: Target lesions, usually found on the extensor surfaces esp. of peripheries; also palms and soles. Major form: (Stevens–Johnson syndrome) associated with systemic upset, fever, severe mucosal involvement, including conjunctivae. Idiopathic; herpes simplex, mycoplasma, other viruses (minor form); drugs esp. sulfonamides, penicillins (major form). Treat the cause; supportive management—use of oral steroids is controversial and may ↑ mortality. www.bxt.co.uk/full.cfm?1116

Erythema chronicum migrans Early sign of Lyme disease (*Borrelia burgdorferi*). 50% give history of preceding tick bite. Presents as small papule which develops into a spreading erythematous ring. Can persist from weeks to months. eg amoxicillin 500mg/8h po, 3wks doxycycline 100mg/12h po. Other features of Lyme disease: OHCM p 602. www.bxt.co.uk/full.cfm?1107

Cutaneous vasculitis There is a variable picture, eg palpable purpuric rash, often on legs; nodules; ulcers; livedo. May occur as part of systemic vasculitis, eg polyarteritis nodosa, Wegener's granulomatosis, Henoch–Schönlein purpura (vasculitic rash on legs and buttocks associated with arthralgia, abdominal pain ± glomerulonephritis). Vasculitis limited to skin (hypersensitivity/leukocytoclastic vasculitis) may be precipitated by infection (streps, hepatitis B & C viruses); thiazides etc; connective tissue disease; malignancy; cryoglobulinaemia. www.bxt.co.uk/full.cfm?1110

Skin diagnoses not to be missed

Actinic keratosis occur on sun-exposed skin, as pre-malignant crumbly yellow-white scaly crusts $\leq 1\text{mm}$ across, as a result of dysplastic intra-epidermal proliferation of atypical keratinocytes.

40yrs old, 21%; if 70yrs old, 52%. Many regress over a year or so—but 15% recur. Bowen's, psoriasis, seborrhoeic wart, BCC; if doubt, biopsy. OPPOSITE. Education; hats; sunscreen

± ↓ dietary fat—but enough vitamin A. Drug Ther Bul; medline:2186792 &: 103354

Basaloid cystic carcinoma Typically, pearly nodules with rolled telangiectatic edge usually on the face. Metastases are very rare. It is locally destructive if left untreated. Lesions on trunk can appear as red scaly plaques with raised smooth edge. uv exposure.

Excision is best; radiotherapy can be used for larger lesions in the elderly. Cryotherapy ± curettage can be used in non-critical sites. medline:105830

Bowen's disease Slow-growing red scaly plaque, eg on lower leg. Full-thickness dysplasia (carcinoma-in-situ). Infrequently progresses to squamous cell cancer. Cryotherapy; topical 5-fluorouracil as above; photodynamic therapy. www.txt.co.uk/full.cfm?1118 medline:112985

Kaposi's sarcoma (p 599).

Leukoderma Rare; suspect in any hypopigmented anaesthetic skin lesion.

Leukoplakia (p 598).

12 yrs
www.txt.co.uk/full.cfm?11

Melanoma (p 584; PLATE 8.1–8.3).

Melanoma Skin metastases are uncommon but well recognized in association with carcinomas of breast, kidney, and lung. Non-Hodgkin lymphoma and leukaemia can also metastasize to the skin. Metastases are usually firm, rather inflammatory lesions; often on the scalp or trunk.

Mycosis fungoides (Skin CD4 T-cell lymphoma). May be long history of undiagnosed plaques—eg well-defined red scaly plaques on trunk/limbs. Can have protracted course and patients may finally succumb to another unrelated disease. Treatments include potent topical steroids, PUVA (p 587) and electron beam therapy. Leukaemic phase (Sezary syndrome) associated with erythroderma and circulating Sezary cells. medline:927636

Nipple's disease of the nipple is a cutaneous sign of intraductal breast cancer. It presents as a unilateral, red, scaly, or crusted nipple, which is often itchy. Biopsy. The chief differential diagnosis is eczema.

Don't diagnose nipple eczema without at least considering a biopsy.

Mastectomy or lesser surgery ± radiotherapy. medline:11237774 &: 930570

Pyoderma gangrenosum Presents as a persistently ulcerated or crusted firm irregular lesion. Usually found on sun-exposed sites, eg ears, dorsa of hands; bald scalp—here it may develop in a pre-existing actinic keratosis (below). Also related to smoking (on the lower lip) and chronic inflammation, eg venous leg ulcers. Excision. Metastases are relatively rare, but more frequent at some sites, eg the ears. www.txt.co.uk/full.cfm?112

Syphilis Isolated painless genital ulcers (primary chancre). Pityriasis rosea like copper-coloured rashes affecting trunk, limbs, palms, and soles (2° syphilis). Are other sexually transmitted diseases present? www.txt.co.uk/full.cfm?112

Options in solar keratosis (None is of proven superiority)

Cryotherapy. After any cryo warn that blisters may form afterwards; they may be burst, repeatedly, with a sterile needle; advise using an anti-septic cream to prevent infection, and aspirin for pain, in adults.

Daily 5% *5-FU* (fluorouracil) cream, with this sequence of events: erythema→vesiculation→erosion→ulceration (red and sore)→necrosis→healing epithelialization. This inflammatory response is in pre-actinic lesions: healthy skin is unharmed. Using pulsed therapy (eg with gaps of a week or more) reduces this inflammatory problem.

3% *diclofenac* gel (Solaraze®) is effective (mechanism unknown); used twice daily for 2 months, it is well tolerated. medline:11841372 & medline11737438

5% *imiquimod* 3 times a week for 6 weeks may be tried to enhance cell mediated immunity via induction of interferon- α . medline:11359396 & 2636692

Moles and melanomas

PLATE 8.1-8.

Melanoma melanoma ♀/♂ 1.3:1. $\geq 10/100,000/\text{yr}$ (has risen in the last 20yrs by ~300% in men and ~200% in women). 5-y survival is rising (now >80%). ~4% have a family history of melanoma.

1.9/100,000/yr in UK. <http://image.thelancet.com/extras/01art7335web.pdf>

Excessive uv exposure. 66% are related to BRAF gene missense mutations (T→A 'mis-spelling'). <http://bmj.com/cgi/content/full/324/7351/1412>

Most arise in normal skin, a few in benign or dysplastic naevi. 'The Glasgow 7': major—Change in: 1 Size (in either vertical or horizontal plane) 2 Shape 3 Colour. Minor: 4 Diameter >7mm 5 Inflammation 6 Oozing/bleeding 7 Itch/odd sensation. Always have a high index of suspicion for changing moles, particularly if the edge is irregular and the pigmentation varies in any mole. ▶ Any lesion with one major feature should be considered for excision. Any minor features add to the suspicion. Differential diagnosis: benign melanocytic lesions; non-melanocytic pigmented lesions (eg seborrheic keratoses—common in those >50 yrs old; they feel greasy, and look as though they have been stuck on). Benign moles tend to be <5mm across, with uniform colour and shape, whereas early melanomas frequently are different shades of brown, black, red, or even blue.

Treatment Always consider excision biopsy with a margin of normal skin and for any unusual, growing or changing pigmented lesion. If histology shows melanoma, read the report carefully: prognosis depends on completeness of excision and tumour thickness (the 'Breslow thickness' is well-established, major prognostic sign). Tumours less than 1mm thick have a very good disease-free 5-year survival rate (>90%), while those thicker than 4mm have only a 50% 5-year disease-free survival rate.*

Melanomas must be completely excised but the margin of excision of normal skin is not yet fully established. A useful working rule is to excise 1cm of normal skin around the melanoma for every millimetre of depth up to 3cm (there is no evidence that excising margins >3cm is of any additional benefit). Enquire if your unit is involved in any of the trials now being done on this issue. *Don't do incisional biopsies on possible melanomas.*

Early diagnosis is vital—everyone should know what an early melanoma looks like, and know how to get prompt expert help. www.txt.co.uk/full.cfm?112

Prevention Evidence suggests that excessive uv light acting on genetically vulnerable white-skinned, fair-haired, blue-eyed individuals is a major cause, so the public should be encouraged to avoid excessive burning sun exposure (noonday sun, eg on sunny holidays), to wear sun-protective clothing such as hats and to use broad-spectrum high SPF (sun protection factor) sunscreens to minimize the risk of burning.

Pigmented naevi or moles It is normal to have benign pigmented naevi or moles on the skin. 3% of us have ≥ 2 congenital pigmented naevi present at birth; numbers slowly increase until by late teenage years, most have 20–50 small naevi. As patients mature, naevi gradually involute so that in old age, benign pigmented naevi are relatively rare on the skin surface. The great majority of naevi are totally benign and are not pre-malignant lesions. Lesions which are less than 5mm in diameter and have a uniform colour and outline do not require to be excised for medical reasons unless there is a clear history of growth or change. www.txt.co.uk/full.cfm?112

*TNM staging is complex, leave to experts, note that melanoma thickness and ulceration but not level of invasion is now used in the T category (except for T1), the number of metastatic lymph nodes rather than gross dimensions is used in the N category, the site of distant metastases and LDH1 are used in the M category. www.txt.co.uk/full.cfm?112

Skin sensitivity to ultraviolet (UV) light

Every individual has a genetically determined sensitivity to ultraviolet radiation. Regardless of race, we all fit into one of 6 skin types.

Skin type

I	Always burns easily, never tans
II	Always burns, tans minimally
III	Sometimes burns, always tans
IV	Rarely burns, always tans well
V	Rarely burns, always tans deeply
VI	Never burns, tans if +++exposure

Those affected

Caucasians (many celtics)
Caucasians
Caucasians
Caucasians
Dark-skinned races (Latin, Indian, Native American)
Afro-Americans

This classification is useful in predicting individual susceptibility for development of skin cancers and manifestations of photoageing. While persons in groups I–II are at highest risk, most skin cancers cluster in type II & III people: type I people know from experience to stay out of the sun. Useful questions in determining type:

When you go out in the sun for the first time each year do you sunburn?

If so, do you always burn? Easily?

Do you ever blister when you burn?

Can you tan? Does it take a long time or do you tan easily?

Do you develop a deep tan or just a shade beyond your normal colour?

See T Fitzpatrick, Arch Dermat 1988;124:869 www.ukpractice.net/portal/medunet/courses

History & exam ▶ Don't just diagnose today's melanoma. You need to prevent tomorrow's by educating the whole family: sun exposure in childhood is a strong determinant of melanoma risk. medline:11134919

Tailor your advice to your patient's risk. Those with highest risk may benefit from professional skin exam every 6–12 months. Would you like information about harmful effects of solar radiation? Ask about family and personal history of freckles/melanomas, and past sun exposure, esp. exposure that caused blistering. Do you use sun-screens? Do you practice safe-skin techniques & skin self-examination? Are your jobs/hobbies outdoor? Are you pregnant? (moles darken during pregnancy). Is hair colour red or blond? Are the eyes blue? Note number and type of all moles (examine all over). NB: melanoma colours are often not all blacks: if melanomas incite inflammation, pink-reds may appear. If the immune response is active, areas of regression with dermal scarring appear as white or greys, depending on residual pigment. medline:2249043 & 3921200 & 2049635

When to refer Think of ABCD: asymmetric lesions, border irregularity, colour irregularity, and diameter >6mm. Refer if ≥3—or if there is a significant spontaneous change in any of A, B, C, or D—or itching, pain, or bleeding.

Sunscreens The SPF (sun protection factor) indicates how much longer an individual can be exposed to sunshine without burning, eg SPF8 means that an individual can stay out in the sun 8 times longer than the time it would normally take them to burn. This time will vary depending on the individual's skin type. The SPF refers only to UVB protection. Many sunscreens now contain UVA-blocking agents. These are graded on a 'star-rating', which as yet, is not internationally standardized.

Sunscreens protect by physically blocking light (physical agents, eg titanium dioxide) or by absorbing light by photochemical reaction (eg benzophenones; cinnamates). Regardless of the mode of action, sunscreens must be regularly applied to all exposed areas (face; back and V of neck; dorsum of hands; legs). Best of all, avoid the sun between 11AM and 3PM, and wear photoprotective clothing to ↓uv exposure.

Psoriasis

PLATE 8.11-8.12

Psoriasis is a common chronic inflammatory skin condition affecting ~2% of Caucasians of any age, peaking in the 20s (type 1) and 50s (type 2). $\sigma/\phi \approx 1:1$.

There are two pathologies: (with abnormal differentiation) and T-cell driven of the dermis and epidermis. The signal to hyperproliferation is (partly) tumour necrosis factor (TNF- α —which is why infliximab, below, is a logical therapy).

30% of patients have a family member with psoriasis. If both parents have psoriasis, the chance of their offspring being affected is ~50%. Genetic linkage studies reveal psoriasis-susceptibility loci on chromosome 6p, 17q & 4q. HLA associations include HLA-CW6, HLA-B13, B17, and DR7. Infections, particularly with streptococci and certain drugs (eg β -blockers; lithium, antimalarials) are also recognized triggers. *Histology*: Epidermal keratinocyte hyperproliferation, parakeratosis, and intraepidermal neutrophil microabscesses (of Munro). medline:9158151

Signs Look for symmetrical well-defined red plaques with silvery scale, on extensor aspects of the elbows, knees, scalp, and sacrum. Flexures (axillae, groins, submammary areas, and umbilicus) also frequently affected but lesions are non-scaly. (in 50%): pitting, onycholysis (separation from nail bed), thickening and subungual hyperkeratosis. Small plaques (guttate) are seen in the young (especially if associated with concurrent streptococcal infection). www.bxt.co.uk/full.cfm?1101 (sterile) can affect the palms & soles. www.bxt.co.uk/full.cfm?1102 (and generalized pustular psoriasis) may cause severe systemic upset (fever, \uparrow WCC, dehydration)—also triggered by rapid withdrawal of systemic steroids.

Other signs Köbner phenomenon (p 576); Auspitz sign: pinpoint bleeding when scale removed; pepper pot nail pitting and 'grease-spots'.

Systemic signs 7% develop arthropathy, which can be of 5 types

- 1 Asymmetrical oligomonoarthritis www.bxt.co.uk/full.cfm?1113
- 2 Predominant DIP joints
- 3 Rheumatoid-like polyarthritis (sero-ve)
- 4 Arthritis mutilans (severe, destructive)
- 5 Psoriatic spondylitis

Differential diagnosis Eczema; fungal infection (solitary or few lesions; asymmetrical; expanding); mycosis fungoides (asymmetrical lesions, minimal scaling, biopsy required); seborrhoeic dermatitis (may co-exist).

Management Education is vital as control, not cure, is the practical aim. Remove possible triggers (streptococcal infection, drugs); factors such as stress and alcohol may also aggravate condition. Topical drugs are the mainstays of treatment for most patients. *Tar*: Messy ointments are best avoided unless patients are admitted for treatment; 'clean' preparations are available (eg Alphosyl[®], applied twice daily). *Dithranol*: Available in creams for use in short-contact regimens (applied carefully to affected skin then washed off after 20–30min): start at low concentrations (0.1%) and increase as tolerated (eg 0.25%, 0.5%, 1%). *SE*: burning (avoid in flexures); staining (avoid on face). *Calcipotriol/tacalcitol*: (vit. D analogues): avoid use in calcium metabolism disorders; small risk of hypercalcaemia if large amounts used on extensive inflamed skin (use calcipotriol up to twice-daily; max 75g per week). Tacalcitol may be applied once daily before going to bed (max 5g/day, and only up to two 12-week courses/yr). For flexural disease, topical steroid/antibiotic/antifungal preparations can be useful (*Trimovate*[®]). *MTX*: Methotrexate helps psoriatic arthropathy. medline:10908464



Plate 8.1 Malignant melanoma. Note the irregular pigmentation and irregular edge.



Plate 8.2 Malignant melanoma. Note the irregular and variable dark pigmentation and the irregular edge. The upper area is nodular and ulcerated.

The images and their legends are supplied by Dr Susannah Baron, whom we sincerely thank.



Plate 8.3 Acral malignant melanoma. This type of melanoma is found on the palms and soles of the feet. Melanomas on the soles of the feet are often not noticed by patients until they are quite large.



Plate 8.4 Lentigo maligna. This is the precursor to invasive lentigo maligna melanoma. It develops in chronically sun damaged skin. Note the lesion is flat and has variable pigmentation and an irregular edge. The darkly pigmented area may be invasive.

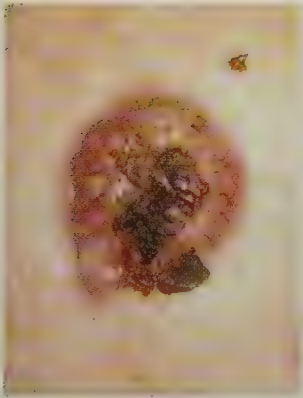


Plate 8.5 Basal cell carcinoma (rodent ulcer). This is a nodular BCC. Note the rolled pearly edge with central ulceration and crust.



Plate 8.6. Basal cell carcinoma. This BCC has extended over the nose. Note the rolled edge, telangiectasia, crusted central ulceration and second smaller cystic BCC by the right inner canthus.

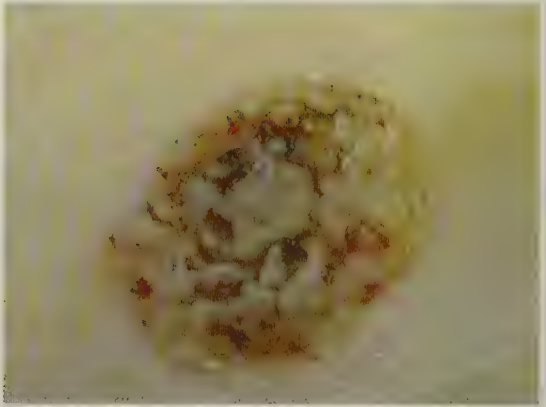


Plate 8.7 Basal cell papilloma (seborrheic wart). This is a benign lesion. It is well defined and has a classic warty stuck on appearance.



Plate 8.8 Atopic eczema. This type of eczema is classically flexural. Note the erythema and excoriations.

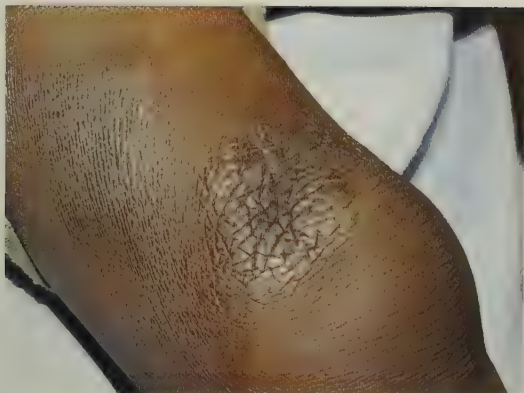


Plate 8.9 Lichenified eczema. This is due to chronic scratching and rubbing. Note that the skin creases are thickened. The darker areas are due to post inflammatory



Plate 8.10 Lichenified infected flexural eczema. Note the skin thickening (lichenification) from chronic eczema, the yellow crust and underlying excoriation, oedema and erythema. Secondary bacterial infection is commonly due to *Staphylococcus aureus*.



Plate 8.11 Psoriasis. This is a plaque of chronic psoriasis on the knee. Note it is well defined, erythematous (salmon pink) and covered with a silver scale.



Plate 8.12 Psoriasis. This is a mixture of guttate and plaque psoriasis. The plaques have occurred where the lesions have become confluent.



Plate 8.13 Scalp psoriasis. Note the red scaly plaques which are well defined and extend beyond the hairline.



Plate 8.14 Venous leg ulcer. This typically occurs over the medial malleolus. Note the yellow slough and surrounding varicose eczema.



Plate 8.15 Widespread drug eruption. It is a macular papular erythematous eruption, confluent in areas with some targetoid lesions.



Plate 8.16 Toxic epidermal necrolysis: This is a dermatological emergency. Most commonly caused by a drug reaction (eg sulphonamides, anti-convulsants, NSAIDs). Note the ulceration of the mucous membranes. Involvement of the eyes, mouth and genitalia often occurs

Agents to consider in recalcitrant psoriasis

Tazarotene (a topical retinoid): for mild to moderate psoriasis affecting <10% skin surface; applied once daily; avoid use in pregnant women.

Phototherapy: Narrow-band UVB (TL-01): 3 × weekly for 6 weeks; avoid if history of photosensitivity; most suitable for guttate/small plaque psoriasis. **PUVA:** UVA + oral/topical psoralen; suitable for extensive large plaque disease (oral psoralen) and localized psoriasis (topical psoralen); limit total dose to 1000J/150 treatments to avoid excessive skin ageing and risk of skin cancer (especially squamous cell carcinoma); can be combined with oral retinoids (re-PUVA) to ↓light dose needed to clear lesions.

Oral drugs: Severe psoriasis often needs oral drugs. Liaise with an expert.

Methotrexate: 10–25mg/week; most useful in elderly patients; best avoided in younger patients in view of long-term risk of hepatic fibrosis.

Ciclosporin: 2.5–5mg/kg/day; usefulness often limited by side-effects (hypertension; renal dysfunction).

Acitretin: Oral retinoid; useful for moderate to severe disease; side-effects: teratogenic; dry skin and mucosae; ↑lipids, ↑LFTs (reversible). In the UK, use is limited to hospitals. Starting dose: 25–30mg/24h po; typical maintenance dose: 25–50mg/day; adjusted according to response). Exclude pregnancy, and avoid donating blood for >1yr and pregnancy until >2yr after the last dose.

Hydroxycarbamide (=hydroxyurea): 0.5–1.5g/24h po. Main SE: marrow suppression.

2001 medline:11703528

iv drugs: **Infliximab** may achieve a high degree of benefit in moderate and severe psoriasis. Initial small studies need verification. Infliximab is a TNF-α/avid monoclonal antibody.

N=31
medline:11410193

Eczema (dermatitis)

PLATE 8.8-8.9

Eczema may be atopic (the commonest form), allergic (type VI hypersensitivity), or be caused by irritants or venous stasis. See also discoid eczema (p 578). Different types may co-exist. Remember to ask about work, hobbies, and other reasons for exposure to allergens.

150 images
www.txt.co.uk/full.cfm?110

Atopic eczema Causes: multifactorial: **Genetic** a family history of atopy common (70%); Overactive TH2 lymphocytes produce IL-4 & IL-5, stimulating IgE production. **Infection** Staph aureus colonises lesions and toxins act as superantigen. **Allergens** ↑IgE is common. RAST (radioallergosorbent testing) identifies specific antigens, eg house dust mite or animal dander. **Diet** Some atopic children have significant food allergies, eg dairy products ± egg which can exacerbate eczema. Although infantile eczema is common (~2% of infants), most grow out of it before 13 years of age. **Treatment**

- Explain that management involves control, not cure. Patients will have dry skin (xerosis), which itches and is susceptible to irritant substances, so emollients are essential as they treat dryness and act as a barrier.
- Bath emollients counteract the drying effects of bathing and some contain antiseptics. Use soap substitutes such as emulsifying ointment.
- Emollients (use at least twice a day). In severe eczema, use of greasy emollients are best, eg emulsifying ointment, eg 50/50 emulsifying ointment and liquid paraffin (~£3/week); epaderm® costs ~£7/week. In less severe eczema, less greasy preparations are more acceptable, eg Diprobase® (~£7/week).
- Topical steroids to areas of active eczema, usually twice daily. Being greasy ointments are better than creams. Strength of steroid depends on severity, site, and age of the patient. Use 1–2.5% hydrocortisone on face and flexures. Elsewhere, get control within 7 days with a higher-potency steroid (eg betamethasone 0.1% ointment), reducing strength and frequency to maintain control (eg betamethasone 0.025%). Clobetasol is an example of an even higher potency steroid. Overuse/abuse of topical steroids in children can lead to skin thinning, telangiectasia ± adrenal suppression.

Irritant dermatitis We are all susceptible to irritants. Hands are often affected; redness ± weeping precedes dry fissuring. **Common irritants**: detergents, soaps, oils, solvents, alkalis; water (if repeated). It often affects bar staff and cleaners. **Treatment**. Avoid all irritants if possible. Good hand care (soap substitutes; regular use of emollients; careful drying of hands after washing; use of cotton or cotton-lined rubber gloves for dry and wet work respectively; intermittent use of topical steroids for acute flare-ups).

Allergic contact dermatitis Cutaneous type IV reaction. Common allergens include nickel (jewellery, watches, coins, keys etc.); chromate (cements, leather); lanolin (creams, cosmetics); colophony (sticking plasters, glues, inks); rubber (foam in furniture); plants (commonly primulas); topical antibiotics (neomycin, framycetin); topical antihistamines and topical anaesthetics (haemorrhoid creams). The pattern of contact often gives a clue to the allergen. There is often sharp cut-off where contact ends but secondary spread elsewhere is frequent (autosensitization). **Treatment**. Consider patch testing and avoidance of implicated allergens; topical steroid appropriate for severity (↓ and stop as the condition settles).

Adult seborrhoeic dermatitis This common red, scaly eruption affects scalp (dandruff), eyebrows, nasolabial folds, cheeks, and flexures. Cause: overgrowth of yeasts (*pityrosporum ovale*) in skin. Can be very severe in predisposed individuals. **Treatment**. Mild topical steroid/anti-fungal preparations eg Daktacort®; ketoconazole cream, lithium succinate ointment. Treatments required intermittently depending on disease activity.

Helping children with atopic eczema*

- Dry skin is itchy, so scratching is a big problem; it may become a habit, so distraction therapies can help, and star charts act as encouragement.
- Dermatology nurses play a big rôle in providing practical support for parents ('he will grow out of it: it won't be like this forever'). They can show how to apply treatments and occlusive dressings, eg stockingette suits, which aid emollient absorption, and prevent drying and scratching.
- Turn down the central heating (it is very drying).
- Sedating antihistamines may be helpful, eg hydroxyzine or alimemazine. High doses may be needed in breaking the itch-scratch cycle, and in enabling the whole family to get a good night's sleep.
- Encourage joining a national eczema association (in UK, 020 7388 4097).
- Discourage elimination diets. Only occasionally is there a clear trigger (eg confirm a dairy allergy with a RAST test; if +ve, get a dietician's help).
- Reducing exposure to house dust mite may help (high filtration vacuuming of mattresses, limiting of carpet use; gortex mattress covers).

The future: nonsteroidal immunomodulation Tacrolimus 0.03% ointment **may** have a rôle in those >2yrs old, used twice-daily on any skin, including face & flexures, for ≤3 weeks (usually by a dermatologist). Stop if no effect after 2 weeks. Long-term safety is unknown, but apparently it has no potential to cause skin atrophy. One advantage is an ability to produce benefits even after treatment ceases. medline:11866675 & 11898004 & 11952659

Preventing infantile eczema

44 .ma
www.txt.co.uk/full.cfm?1100

Despite the numerous studies on possible benefits of breastfeeding, the issue remains controversial: one meta-analysis suggests that exclusive breastfeeding for the 1st 3 months of life is associated with less atopic eczema in children with a 1st degree relative with atopy. medline:11568741

Prescribing antigen avoidance diets to high-risk women during pregnancy has little effect medline:10796150 but during lactation this *might** 'substantially reduce her child's risk of developing atopic eczema, but better trials are needed'. If a mother is keen to try antigen avoidance, point out that it may not work; at present there is not enough evidence for us to be persuading those who are neutral on the issue. medline:10796149

- If breastfeeding is impossible, many try soya milk (eg Wysoy®)—but this does not work. The most meticulous blind trials show that a much better alternative is a milk in which the protein (casein) is hydrolysed (eg Nutramigen® or Pregestimil®). medline:10981524

- Reducing exposure to house-dust mite: www.ful-text.com/r.cfm?1013 *ebm*
High-filtration vacuuming of mattresses Acaricidal sprays, eg benzyltannate Gore-tex® covering of mattresses Washing bedding at 55°C

NB: some meta-analyses say that mite control is unproven. medline:11134919

Williams diagnostic criteria for atopic eczema medline:11531832

A child must have an itchy skin (or a parental report of scratching) plus 3 or more of:

- Onset before 2yrs old (this criterion is not used if child <4yrs old).
- History of skin crease involvement (including cheeks if <10yrs old)
- History of generally dry skin.
- Personal history of other atopy or history of atopy in 1st degree relative.
- Visible flexural dermatitis (or dermatitis on cheeks or forehead and outer aspect of limbs if <4yrs).

*See NHS HTA systematic review www.nccta.org & Cochrane reviews [a10796150] [a10796148]

Skin infections

www.pediatrics.wisc.edu/derm/tuttc/50.html#

Fungi The superficial mycoses are the commonest of human fungal infections—and are limited to skin, hair, nails, and mucous membranes. They include dermatophytes (ringworm), superficial candidosis, and Pityriasis sporum infections. Three genera of dermatophytes affect human: Trichophyton, Microsporum, and Epidermophyton and can be spread from man to man (anthropophilic) eg *T rubrum*, animal to man (zoophilic) (eg *M canis*) or soil to man (geophilic), eg *M gypseum*. A dermatophyte infection is a round, scaly, itchy lesion whose edge is more inflamed than its centre. It is called tinea followed, in Latin, by the part affected, eg tinea pedis (foot); cruris (groins); capitis (scalp); unguium (nail); corporis (body). Infection caused by yeasts of the Candida genus (eg *C albicans*) are common particularly in the immunocompromised (colonization→invasion→dissemination, *OHCM* p 614–5). Skin infections are often pink and moist, and there may be satellite lesions. Sites involved by candida: mouth, vagina, glans, skin folds/toe web; nail areas. Other yeasts causing infection include *Pityrosporum ovale* and *orbiculare* in seborrhoeic dermatitis, and *Malassezia furfur* in Pityriasis versicolor (multiple hypo- or hyperpigmented scaly macules on the upper trunk and back). **Diagnosis.** Skin scrapings are taken by scraping from the edge (active margin) of the lesion, with a blunted scalpel. Hair-pulls and nail clippings should be taken and specimens collected in folds of black paper (contrasting with the white scrapings). A Wood's light which causes fluorescence in some fungal infections can help diagnosis, and suggest areas for scrapings. Microscopy (potassium hydroxide added to the scrapings helps dissolve keratin) and culture (takes 6 weeks) also help. Skin swabs should be taken if candidosis is suspected. **Treatment.** *Dermatophyte infections:* Skin—terbinafine or imidazole creams (eg clotrimazole) applied twice daily for 14 days. Scalp—oral griseofulvin or terbinafine for children for 2 weeks. Nails—oral terbinafine for 3 months. If treatment is desired, after explaining about side-effects (headache; dizziness; taste disturbance; arthralgia; liver failure; psychiatric illness; vertigo) and interactions (β -blockers; antidepressants).

Candida: Mouth—nystatin (eg pastilles, sucked 4 times a day for 1 week). Vagina—imidazole cream \pm pessary (eg Gyno-Daktarin Combipack®).

Pityriasis versicolor: Selenium sulfide shampoo applied at night, and washed off in the morning, or topical imidazole creams used twice daily.

Bacteria *Impetigo* Superficial *Staph aureus* infection, common in children and often starting around the nose and face. **Clinical features:** Honey-coloured crusts on erythematous base; lesions often well-defined and superficial flaccid blisters sometimes seen. **Treatment:** Topical fusidic acid or systemic antibiotics (eg flucloxacillin 125mg/6h po, in a child).

Erysipelas Sharply defined superficial infection caused by streptococcus. Often affects the face (unilateral). Associated with fever, leukocytosis. **Treatment:** Systemic penicillin (see below).

Cellulitis. Deeper streptococcal infection with systemic upset & lymphadenopathy, less well-defined than erysipelas and often affecting legs. Commonest organisms: β -haemolytic streps \pm staphs. **Treatment:** \blacktriangleright Elevate the legs. Benzylpenicillin 600mg/6h iv (or penicillin V 250–500mg/4–6h po) + flucloxacillin 500mg/6h po (if penicillin-allergic: erythromycin 500mg/12h po).

Common viral infections of the skin

Warts Cause: human papillomavirus (HPV) infection of keratinocytes. Large numbers of lesions often seen in the immunosuppressed, eg transplant patients. *Common warts* Most common in children and young adults; often resolve spontaneously but individual lesions can be stubbornly persistent. Treatment is either expectant (wait-and-see) or destructive: topical salicylic acid paints (keratolytic); liquid nitrogen cryotherapy (if the child is >6–8yrs old); stubborn lesions are occasionally treated with intralesional bleomycin ± lasers. 28 ima www.txt.co.uk/full.cfm?1097

Plantar warts: Large confluent lesions (mosaic warts) are often resistant to repeated treatments. Warts are infectious and excision is best avoided. After abrading the wart, salicylic acid (eg 26%) is sometimes tried, under an occlusive plaster, eg as Occlusal®. Cryo: double freeze-thaw cycles are more effective than single cycles (but not for hand warts). medline:7857844

Schools expect children to wear verruca socks for swimming. One uncontrolled study somewhat supports this: public shower users were at greater risk for plantar warts than were locker room users who did not use communal showers. medline:7852935 www.txt.co.uk/full.cfm?1096

Plane warts: Flat skin-coloured or brown lesions; tend to Köbnerise (p 576) in scratch marks; they often resist treatment. www.txt.co.uk/full.cfm?1095

Genital warts (condylomata acuminata): Usually treated with cryo ± podophyllin (avoid in pregnancy: teratogenic) or imiquimod cream. Also screen for other sexually transmitted diseases. Women with genital warts (or whose partners have them) need a yearly cervical smear (HPV 16 & 18 contribute to cervical cancer, p 34). www.txt.co.uk/full.cfm?1094

Molluscum contagiosum (pox virus): These pink papules have umbilicated central punctum. White material can be expressed and microscopied to confirm the diagnosis (molluscum bodies). Common in children. They resolve spontaneously (but this may take months). Gentle cryo may be tried. It is more common in atopic eczema. 8 image www.txt.co.uk/full.cfm?1113

Herpes simplex Primary infection is often subclinical. Most frequent clinical manifestation is gingivostomatitis. *Recurrent infection:* Is commonly genital or perioral. Triggered by fevers, sunlight, immunosuppression. Eruption often preceded by symptoms of burning/itching. *Signs:* Grouped painful vesicles on erythematous base which heal without scarring. *Treatment:* Often none required. Topical aciclovir may prevent or reduce severity of recurrences. Systemic treatment is indicated in certain circumstances (immunosuppressed; frequent recurrent genital herpes). There is evidence that Bell's palsy may be related to recurrent herpes infection. 4 image www.txt.co.uk/full.cfm?1098

Herpes zoster (shingles): Primary infection is chickenpox (p 216). Varicella-zoster virus becomes dormant in dorsal root ganglia. Recurrent infection affects one or more dermatomes (esp. immunosuppressed, when it may become generalized). Pain and malaise may precede eruption. *Signs:* Polymorphic eruption (red papules, vesicles pustules, and crusting). May heal with scarring and development of post-herpetic neuralgia. *Rx:* If mild, none may be needed. More severe disease or infection in immunosuppressed requires early treatment with systemic treatment, eg aciclovir 800mg 5x/day po for 7 days (SE: confusion; LFT↑; urea↑)—or famciclovir 250mg/8h po for 7 days (SE: headache, confusion). 32 ima www.txt.co.uk/full.cfm?1099

Acne vulgaris

This is an inflammatory disorder of the pilosebaceous follicle. It is almost universal amongst teenagers, affecting the face, neck, upper chest, and back.

Cause Comedone (whitehead & blackhead) formation is the central abnormality in acne and is due to an abnormality of keratinization/desquamation within the follicle, leading to a blockage of secretions. * *Sebum production* This is regulated by androgens and CRH → see p 575. Most individuals with acne do not have excessive androgens but their sebaceous glands may be more sensitive to the actions of androgens. *Propionibacterium acnes* *P. acnes* is a normal skin commensal that flourishes in the anaerobic conditions within the blocked follicle. It is the inflammatory response to this bacterial proliferation which causes the typical inflamed lesions of acne.

Signs (Face, trunk) Comedones; papules; pustules; nodules; cysts; scars.

Differential diagnosis Acne rosacea—no comedones; diffusely red nose, cheeks, chin, and forehead; telangiectasia; flushing, especially after alcohol.

Management Dispel some of the myths about acne—it is NOT due to lack of washing, eating sweets/chocolate, etc. Treatment will depend on severity.

Mild acne (mainly comedones, mainly on the face): topical therapies (monotherapy or in combination): benzoyl peroxide (as low a strength as is effective, eg 2.5%; if higher, it may irritate); azelaic acid (max 10g/day; treat for no longer than 6 months); roll-on antibiotics (clindamycin as Dalacin T[®]); retinoids.

Moderate acne (mainly inflammatory lesions, face ± chest, back): oral antibiotics are the mainstay of treatment—eg modified-release minocycline 100mg/day po; erythromycin 500mg/12h po. Courses should be 4–6 months minimum. To avoid development of bacterial resistance, benzoyl peroxide should be used along with oral antibiotics. If topical antibiotic used, this should be the same drug as is being given orally, to avoid multiple drug resistance. **Severe acne** (nodules, cysts, scarring): the synthetic retinoid isotretinoin is the drug of choice. It is usually given for 16 weeks at a dose of 0.5–1mg/kg/day. Highly significant improvements are seen in virtually all patients. 60–70% will have no further recurrences. **SE:** teratogenic (sexually active women must use an effective form of contraception during and for 1 month after treatment); dryness of skin, lips, mucosae, myalgia, headache (benign intracranial hypertension reported); hepatitis; ↑lipids. medline:1127973

Other possible indications for the use of isotretinoin now includes moderate acne which fails to respond to adequate course of antibiotics, the presence of scarring and severe psychological problems.

Acne rosacea

A chronic relapsing and remitting facial disorder of unknown aetiology, particularly in fair-skinned individuals. Chronic flushing triggered by alcohol, spicy foods etc. often precedes the development of fixed erythema (chin, nose, cheeks, forehead), telangiectasia, papules, and pustules. In males, rhinophyma (swelling and soft tissue overgrowth of the nose) may occur. Recurrent severe flares can lead to the development of lymphoedema. Eye problems commonly associated—blepharitis (scaling and irritation at base of eyelashes); conjunctivitis; rarely, keratitis. **Management:** Avoid irritants, excessive sun exposure. Topical metronidazole can effectively control mild to moderate disease. Oral tetracyclines are the mainstay, eg oxytetracycline 500mg/12h po, in an acne type regimen. Once skin has settled, discontinue, and restart as needed (**NB:** long-term use of minocycline can cause blue/grey pigmentation on exposed sites and, very rarely, hepatitis). medline:1079685

10% of hospital patients develops a drug eruption during their admission, so it is important to know which drugs typically cause problems, and to understand how to manage these problems. Drug reactions can be classified according to various criteria, eg immunological (IgE type I response to penicillin) or non-immunological (teratogenicity from retinoids); histological features, eg lichenoid; clinical features.

Types of clinical reaction *Maculopapular exanthematous* This is the most frequent type of cutaneous drug eruption. **Clinical features:** Macular erythema and red papules, particularly affecting trunk. There may be associated fever and eosinophilia. **Drugs:** Penicillins; cephalosporins; anti-epileptics.

Urticaria: **Signs:** Raised itchy red weals with pale centres; rapid onset after taking drug ± association with angio-oedema/anaphylaxis. This can result from both immunological and non-immunological mechanisms. **Drugs:** Morphine, codeine, and other opioids cause direct mast cell degranulation; penicillins and cephalosporins trigger IgE responses; NSAIDs; ACE inhibitors.

Exfoliative dermatitis: **Signs:** Widespread erythema and dermatitis; erythroderma. **Drugs:** Sulfonamides; allopurinol; carbamazepine; gold.

Erythema multiforme major (Stevens–Johnson syndrome): The more severe variants of erythema multiforme are usually due to drugs. **Signs:** Target lesions and polymorphic erythema, sometimes blistering; mucosae (conjunctivae, oral and labial, genital) severely affected. **Drugs:** Sulfonamides; anticonvulsants.

Toxic epidermal necrolysis. (PLATE 8.16) This may represent the extreme end of the erythema multiforme/Stevens–Johnson syndrome spectrum. **Signs:** Widespread erythema followed by epidermal necrosis with loss of large sheets of epidermis. Mucosae severely affected. This has a high mortality rate (>30%). This is best managed in specialist dermatology or burns unit. **Drugs:** Sulfonamides; anticonvulsants; penicillins; allopurinol; NSAIDs.

Lichenoid: **Clinical features:** There are clinical features which show some similarity to lichen planus but are rarely typical of idiopathic lichen planus. **Drugs implicated:** β -blockers; thiazides; gold; antimalarials.

Management A clear history of the onset and duration of the rash is essential. A previous history of skin problems or cutaneous drug reactions should be sought. All drugs taken (currently or previously) should be recorded, including over-the-counter preparations (laxatives, analgesics, herbal remedies, etc.). If the clinical findings are suggestive of a drug eruption, the rash should be classified clinically and the most likely offender identified and discontinued immediately, if possible. If the clinical diagnosis is in doubt, a prick test or skin biopsy may be helpful but is not always so. In order to confirm the suspicion of drug sensitivity, some advocate rechallenge with the suspected drug once the patient has recovered. Whilst this is the ideal situation, rechallenge may be dangerous (eg anaphylaxis, erythroderma). Not unreasonably, patients may also object to this. medline:9582146

Many drug rashes need no intervention. Symptoms such as dryness or itch may be helped by regular emollients. Very itchy rashes, eg lichenoid or dermatitic, may need short courses of topical steroids. Urticaria: prompt antihistamine with iv hydrocortisone/im epinephrine (=adrenaline, p 309) if anaphylaxis. More severe eruptions, eg erythema multiforme are best managed by specialists. There is debate as to the efficacy of oral prednisolone in both of these conditions. As these drugs can potentially adversely affect outcome (eg ↑ risk of infection, hypertension, fluid retention, diabetes, etc.) their use should be determined by specialists.

Miscellaneous skin disorders

Lichen planus This is an itchy disorder (cause unknown), occurring at any age (uncommon in the very young and old). The eruption consists of flat-topped purple papules with white lacy markings on the surface (known as Wickhams striae). The flexor aspects of the wrists, forearm, ankles and legs are commonly affected. Typical lesions at other sites include scalp (scarring alopecia), nails (longitudinal ridges), mouth (lacy white areas on inner cheeks), and genitals (annular lesions). Lesions often arise at sites of trauma. Usually persists for 6–18 months. Treatment symptomatic: moderate to potent topical steroids are helpful for severe itch. Occasionally, severe disease warrants the use of oral steroids.

Haemangiomas Strawberry naevi occur in neonates, as a rapidly enlarging red spot. Most go by the age of 5–7 years. No treatment is required unless a vital function is impaired, eg obscuring vision.

Pyriform granuloma This is a lesion which is thought to arise as a result of minor trauma, typically occurring on fingers. It appears as a moist red lesion which grows rapidly and often bleeds easily. **Treatment:** Curettage.

Pityriasis rosea This is common, often affecting young adults, and may have a viral cause (eg herpes hominis viruses HHV-6 & HHV-7).^{medline:1106945} A rash is preceded by a herald patch (ovoid red scaly patch with prominent scaly edge, similar to but larger than later lesions). Tends to affect the neck, trunk, and proximal limbs. It is self-limiting; no treatment is needed. www.dermis.net/doi/a/diagnose.asp?zugn=d&lang=e&diagnr=694410&topic=

Alopecia Hair loss is classified as scarring or non-scarring. Non-scarring causes may be reversible but scarring alopecia results in an irreversible loss. Disorders of the scalp/hair may be manifestations of skin disease elsewhere: look for evidence of this (eg lichen planus, SLE).

Non-scarring alopecia: *nutritional* (iron deficiency; zinc deficiency rarely); *androgenic* (men & women); *autoimmune*¹ (alopecia areata: smooth round patches of hair loss on scalp; hairs like exclamation marks are a typical feature; often spontaneously regrows; total scalp hair loss = alopecia totalis; total body hair loss = alopecia universalis—its treatment is difficult: consider topical or intralesional steroids, or minoxidil or dinitrochlorobenzene); *telogen effluvium* (shedding of telogen phase hairs after period of stress, eg childbirth, surgery, severe illness).

Scarring alopecia: *lichen planus*; *discoid lupus erythematosus*; *trauma*

Blistering disorders *Infection* (eg herpes); *insect bites* (eg on legs); *drug* (ACE-i; furosemide); *trauma/friction*; *Zn↓**; *autoimmune blistering disorders* (eg pemphigus)—the commonest autoimmune blistering skin disease of the elderly—due to IgG autoantibodies against basement membrane components (bullous pemphigoid antigens 1 & 2). *Signs of pemphigoid*: Tense blisters on an urticated base. Skin biopsy: (+ve immunofluorescence; linear IgG and C3 along the basement membrane). Rx: clobetasol propionate cream 40g/day is better than oral steroids. ^{medline:1182150}

Pemphigus affects younger people than pemphigoid (>40yrs) and is due to IgG autoantibodies against desmosomal components (desmoglein 1 & 3). *Signs*: Flaccid blisters which rupture easily to leave widespread erosions. The oral mucosa is often affected early. Diagnosis is confirmed by skin biopsy with positive immunofluorescence (intercellular IgG giving 'crazy-paving' effect). *Treatment*: Prednisolone (60–80mg/day po, may be life-long at reduced dose). www.dermis.net/doi/a/diagnose.asp?zugn=d&lang=e&diagnr=694410&topic=

*↓ Zinc causes bullae, diarrhoea, and dermatitis (+ loss of eyebrows & lashes, and failure to thrive in acrodermatitis enteropathica: www.bxt.co.uk/full.cfm?1047) 1 V Randall *Lancet* 2001 **358** 192

Photosensitivity

For classification of skin-type susceptibility to ultraviolet, see p 584–5.

Photosensitivity denotes conditions triggered by exposure to light (eg solar urticaria; polymorphic light eruption). Photoaggravation describes disorders worsened by light but are not due to abnormal sensitivity to light (eg recurrent herpes labialis and rosacea). Photosensitivity can be examined by assessing erythematous response to light of a certain wavelength and comparing responses to the normal range. Photosensitivity can occur to visible light, uva (320–400nm) or uvb (290–320nm).

- Usually a careful history is enough to diagnose photosensitivity:
- A rash every summer, for example, suggests that uv has a rôle.
- How long does the rash take to appear? Within 30min suggests solar urticaria; polymorphic light eruption may take days to appear.
- Itching suggests polymorphic light eruption or solar urticaria.
- Face rashes, including the V of the neck, but sparing periorbital skin.
- The dorsum of the hands and the arms are other typical sites.
- As hands are usually relaxed in the semi-flexed position, skin around DIP joints is less affected than that around the knuckles and PIP joints.
- Is there a family history? If so, refer to genophotodermatologist. Is it xeroderma pigmentosum, Bloom's syndrome, or Cockayne's syndrome?
- Pain (± family history) suggests erythropoietic protoporphyria (sun may hurt so much that a child quickly avoids it, so there is pain but no rash).
- Was the rash related to starting a drug (eg nalidixic acid)? See below.
- Are there papules: this suggests polymorphic light eruption.
- Any blistering or linear patterns where plants scratched the skin (phytophotodermatitis from furocoumarins)? Ask about hobbies and work.

www.dermis.net/doi/image.asp?zgr=d&lang=e&cd=33&nr=12&diagnr=692982

Some photosensitive disorders *Polymorphic light eruption* This is a common idiopathic disorder typically affecting young women. During the spring, following light exposure, itchy red papules, vesicles, and plaques develop on exposed sites, often improving over the summer months due to a phenomenon called 'hardening'. *Treatment* Sun-avoidance; sun-protection (high factor uva + uvb sunscreens); desensitization by phototherapy; strong topical steroids ± brief course of oral prednisolone.

medline:11069465

Porphyria cutanea tarda (The commonest porphyria) The primary abnormality is hypofunction of the liver enzyme uroporphyrinogen decarboxylase. *Causes/triggers*: certain alleles of the haemochromatosis gene; HIV; hepatitis C; alcohol; oestrogen. *Signs* Vesicles/bullae in sun-exposed sites, hypertrichosis, hyperpigmentation, skin fragility, and scarring (milia). *Tests*: ↑LFTs; ↑ferritin; ↑urine, plasma, and faecal porphyrins. *Management*: Remove precipitants; sun-avoidance; sun-protection; regular venesection until ferritin in normal range; chloroquine.

medline:10815515

Systemic lupus erythematosus (OHCN p 410) Light exposure often triggers erythematous rashes, with systemic flare. A variant of lupus, subacute LE, is associated with marked photosensitivity and anti-Ro antibodies.

Drugs are a major cause of photosensitivity, so . . . ► look up details of all drugs taken. Frequent offenders: thiazides (sunburn-like or lichenoid eruptions); tetracyclines/sulfonamides (sunburn-like); tricyclics; phenothiazines (dermatitic eruption); NSAIDs; amiodarone (sunburn-like eruptions). It may take months to settle, after drug withdrawal.

Skin disorders in old age

Leg ulcers (CAH 847) *Leg ulcers* Typically venous hypertension from damaged valves of the deep venous system (eg 2° to DVT). This causes superficial varicosities and skin changes (lipodermatosclerosis = induration, pigmentation and inflammation of the skin of the lower leg). Minimal trauma typically over the medial malleolus causes ulcers. *Management* The only effective treatment is compression, ideally in community leg ulcer clinics. Ulcers heal more quickly with occlusive dressings which absorb exudate and improve comfort. Treat varicose eczema with emollients and a short course of moderate-potent topical steroids. Consider patch testing if unresponsive. Bacterial colonization doesn't require treatment **unless** streptococcal or pseudomonal, or there are clear signs of cellulitis/infection (↑pain and tenderness, fever, erythema or lymphangitis). Four-layer compression bandaging should be applied by trained nursing staff, to produce a pressure of 40mmHg at the ankle. Do Dopplers before this to exclude arterial disease (make sure the ankle-brachial pressure index is >0.8, *OHCM* p 478).¹² Ulcers which do not heal on adequate treatment for 3 months should be investigated further (eg biopsy for malignancy). Once the ulcer has been healed, compression hosiery should be worn. Grade III stockings are the ideal but many find them uncomfortable; grade II is a reasonable compromise.

Other causes Peripheral vascular disease (pale cold leg, shiny atrophic skin, absent pulses); neuropathic ulceration (most commonly due to diabetes and seen on the pressure areas of the foot); pyoderma gangrenosum (rheumatoid arthritis; sickle-cell disease; Crohns/ulcerative colitis; idiopathic); trauma; vasculitis (systemic lupus, rheumatoid arthritis).

Asteatotic eczema This is common in the elderly, and particularly affects the lower legs. The skin is dry and eczema is seen in a 'crazy-paving' pattern. The main treatment is the use of emollients and soap substitutes.

Pruritus is a common complaint in the elderly and there are many causes:

Primary skin disorders Eczema; scabies (can have an atypical clinical appearance in the elderly); pemphigoid and pre-pemphigoid eruptions; asteatotic eczema; generalized xerosis. **Systemic medical conditions** Anaemia; polycythaemia; lymphoma; solid neoplasms; hepatic and renal failure; hypothyroidism and hyperthyroidism; diabetes (candidiasis). In the absence of a primary skin disorder, these should be excluded by appropriate investigations.

Pressure sores If made immobile by age, or CNS problems (stroke, cortical lesions, or multiple sclerosis), uninterrupted pressure on an area of skin may lead to ulceration, and extensive, painful, subcutaneous destruction—eg on the sacrum, heel, or greater trochanter. Malnutrition (esp. protein), arteriopathy, and old age make the condition more likely to occur, particularly if nursing care is wanting. They are a big problem. Cost, UK: >£75 million/yr; USA \$3 billion/yr.¹ A full-thickness sacral sore extends hospital stay by >25 weeks, and costs £26,000. ▶ This should make prevention a central preoccupation, not just on long-stay geriatric wards, but in all acute adult wards—where most pressure sores start.

Staging *Stage 1* Non-blanching erythema over intact skin.
Stage 2 Partial thickness skin loss, eg shallow crater.
Stage 3 Full thickness skin loss, extending into fat.
Stage 4 Destruction of muscle, bone, or tendons.

Prevalence ~7% of inpatients have pressure sores; >70% are over 70yr old. Up to 85% of paraplegic patients have pressure sores.

Complications Osteomyelitis; pyoarthrosis.

Treatment and prevention of pressure sores

- Prevent the condition getting worse (see below).
- Improve nutrition. Insulin if hyperglycaemic.
- Treat systemic infection with antibiotics.
- Dress the area. There is no convincing evidence from randomized trials which favours any one type of dressing.
- Vascular reconstruction, if needed, and if practicable.
- Split thickness skin grafts.
- Neurosensory myocutaneous flap surgery.²

Prevention

- Find an interested, knowledgeable nurse to educate the patient.
- Proper positioning, with regular turning (eg every 2h, see p 726 & p 732, alternating between supine, and right or left lateral position). Use pillows to separate the legs.
- Functional electrical stimulation can prevent sores in paraplegics, by inducing the buttocks to change shape, and by improving blood flow.³
- A good randomized trial ($N=44$ patients aged ~85yrs with hip fracture) showed that a DeCube[®] mattress with removable cubes for provision of rest for pressure points can halve the incidence of pressure sores.^{~4~}

1 C Morrell 1998 *BMJ* i 1487 & A Fletcher *BMJ* 1997 **315** 576 2 R Vohra 1994 *BMJ* ii 853
3 A Fergusson 1992 *Paraplegia* **30** 474 ~4~ A Hofman 1994 *Lancet* **343** 568

Skin disorders in HIV disease

There are many cutaneous manifestations of HIV disease, due to both infectious and non-infectious conditions. With the introduction of the protease inhibitor group of drugs and the advent of early treatment with triple therapy regimens, it is likely that some of the conditions described below will become less common in the HIV positive population.

Infections: HIV positive individuals are more prone not only to infections common in the general population but also to infections with organisms which do not usually cause disease in immunocompetent individuals (opportunistic organisms). In an HIV positive patient, any new lesion should be considered as possibly due to infection with an unusual organism.

HIV Seroconversion is associated with an acute mononucleosis type illness usually accompanied by a non-specific maculopapular eruption affecting the upper trunk, associated with lymphadenopathy, malaise, headache, and fever.

Candida may be severe, disseminated, treatment-resistant, involving the posterior pharynx and oesophagus. **Rx:** Topical nystatin; systemic imidazoles.

Molluscum contagiosum Lesions tend to develop on the face and genitals. Management can be difficult: cryotherapy, topical retinoids, cautery, and curettage may be tried. **Differential diagnosis:** Disseminated cryptococcosis.

Herpes simplex virus infection. This can be increasingly troublesome as HIV progresses. Painful ulcers and erosions develop, particularly around the mouth and genitals. Any ulcerated or eroded area should be considered as HSV until proven otherwise. **Management:** High-dose aciclovir (oral or IV). Aciclovir resistance may develop and foscarnet is an alternate treatment.

Herpes zoster. This may occur early in HIV, with atypical presentation. Complications such as ulceration and post-herpetic neuralgia appear to be more frequent and severe. In advanced disease, disseminated infection occurs. **Treatment:** High-dose aciclovir (IV if systemic disease).

Cryptococcosis. Looks like facial molluscum contagiosum. **Rx:** Fluconazole.

Scabies. Severe variants, eg crusted scabies are more common in advanced HIV disease. Paradoxically, patients may not complain of severe itch. A widespread scaly, crusted eruption occurs (highly infectious). **Treatment:** Permethrin lotion. Ivermectin may also be of benefit (but side-effects may be serious).

Oral hairy leukoplakia (Epstein-Barr virus infection of oral mucosa). Adherent white plaques are seen on the lateral aspects of the tongue. **Treatment:** Systemic aciclovir.

medline:1965185

Inflammatory disorders Despite the immunosuppression associated with HIV disease, inflammatory cutaneous conditions occur frequently.

Syphilis and leishmaniasis. Common in later stages of HIV, it may be widespread and severe. Red scaly patches typically affect hair-bearing areas, eg the nasolabial folds, scalp and flexures. **Rx:** Topical (or systemic) imidazoles.

Psoriasis. Standard therapies (tar; dithranol; calcipotriol; UV). Treatment of HIV will often improve psoriasis.

Prurigo simplex. The cause of this condition is unknown and it occurs frequently as HIV progresses. Itchy follicular papules and pustules affect the face, chest, and back. Treatments are often unsatisfactory but include topical steroids; phototherapy; antihistamines.

Drug eruptions. These occur more commonly in HIV patients and particular culprits include co-trimoxazole (maculopapular eruptions; erythema multiforme; toxic epidermal necrolysis—PLATE 8.16); dapsone; foscarnet (ulceration); zidovudine (nail + mucosal pigmentation).

Neoplasia and HIV

HIV disease is associated with a significantly increased risk of certain types of malignancy, eg lymphoma, cervical carcinoma. Common skin cancers (eg basal cell carcinomas, PLATE 8.5) are also seen with HIV.

Kaposi's sarcoma (KS) Although first identified in HIV +ve homosexuals, there is evidence to suggest that HHV-8 (herpes hominis virus) is associated with KS in both HIV positive and negative populations. The condition most frequently arises in advanced HIV disease. **Signs:** Purplish macules, papules, nodules, and plaques affecting the feet, legs, face, and oral mucosa. **Differential:** Bacillary angiomatosis (*Bartonella* species); pyogenic granuloma. **Treatment:** Cryotherapy; intralesional vincristine; radiotherapy; treatment of HIV disease (protease inhibitors).

Infestations

Scabies (*Sarcoptes scabiei*) This is a common disorder particularly affecting children and young adults, and tends to spread within families or those living in close contact. The ♀ mite digs a burrow and lays eggs which hatch as larvae. The itch and subsequent rash is probably due to allergic sensitivity to the mite or its products.

Markedly itchy papular eruption affecting the fingerwebs (esp. first), wrist flexures, axillae, abdomen (esp. around umbilicus and waistband area), buttocks, and groins (itchy red penile or scrotal papules are virtually diagnostic). In young infants, the palms and soles are characteristically involved. The eruption is usually excoriated and becomes eczematized. Scabies mites can sometimes be extracted from burrows and visualized microscopically; eggs can be visualized from skin scraping.

Management: A good explanation (verbal + written) will aid concordance between the patient's and the doctor's requirements, and will promote the chances of successful cure. A suitable anti-scabetic should be prescribed (malathion, permethrin). Preparations are applied to all areas of the skin from the neck down for 24h. Areas which are washed during this period (e.g. hands) should have treatment re-applied. **ALL** members of a household should be treated at the same time, even if asymptomatic. The rash and symptom of itch will take a few weeks to settle, occasionally longer. A suitable anti-pruritic such as crotamiton cream (which also has anti-scabetic activity) can be useful during this period. *Example of written advice:*

- Take a warm bath and soap the skin all over.
- Scrub the fingers and nails with a firm brush. Dry your body.
- Apply malathion 0.5% liquid (Derbac-M[®], not if pregnant or <6 months old) from the neck down (include the head in those <2yrs old). Remember to paint all parts, including the soles.
- Wash off after 24h. If you wash your hands before 24h is up, reapply the liquid to the parts you have washed.
- Use fresh pillow cases and sheets, if you have any.
- Treatment may worsen itch for 2 weeks—so use calamine lotion.
- Warnings: avoid the eyes.

Note: one or two doses of ivermectin 200µg/kg po may be effective in recalcitrant scabies (eg with HIV).

Headlice (*Pediculus humanus capitis*) This is a common problem among schoolchildren, with lice being spread by head to head contact. The louse is 3mm long and has legs adapted to cling onto hair shafts. The eggs (nits) are bound firmly to the scalp hairs and when empty appear white.

Features: Itch and a papular eruption on the nape of the neck are common. **Management:** Resistance to treatment has become a problem and many health boards operate a rotation policy of treatments. Sometimes the only option is meticulous combing (after applying conditioner to the hair).

www.dermis.net/doiia/diagnose.asp?zogr=d&lang=e&diagnr=694410&topic=

Crab lice (*Phthiriasis pubis*) These are usually sexually transmitted and affect the pubic hairs. The eyebrows, eyelashes, and axillae may also be involved. **Management:** Topical malathion to all affected (or potentially affected) areas. Evidence of other sexually transmitted disease should be sought. All sexual contacts should also be treated.

Flea bites (*Pulicidae*) spread plague, typhus, cat-scratch disease. The animal (eg cat or dog) which spreads the flea may not itch or scratch itself. Flea bites cause a papular urticaria in a sensitized individual. Treatment: de-flea pets; de-flea household carpets and soft-furnishings.

Treating head lice

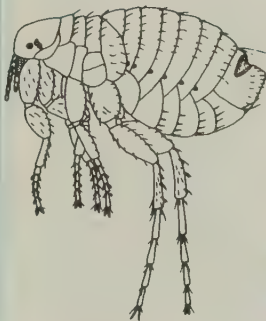
- Only treat living, moving, lice.
- Treatment courses entail 2 applications of >50mL of lotion, 1 week apart.
- Do contact tracing; treat confirmed infections with the same agent.
- In choosing which agent, use the 'mosaic model', to avoid prolonged use of the same agent. Be guided by local patterns of resistance. Try malathion alcoholic lotion (aqueous if young or atopic), then permethrin, then carbaryl (only the latter needs a GP prescription in UK: it is a potential carcinogen). Allow to dry and wash off after 12h, and wet-comb.
- Shampoo formulations rarely work.
- Families should do wet hair 'detection combing' 2–3 days after the 2nd application, to check on cure.
- Don't use insecticides as prophylaxis.
- Prophylaxis: in typical 'cases' (urban girl aged 4–11) regular combing with early intervention may work.
- If drugs fail, use mechanical methods.

MeReC 1999 10 17

The flea

*Mark but this flea, and mark in this
How little that which thou deny'st me is;
Me it suck'd first, and now sucks thee,
And in this flea our two bloods mingled be;
Confess it; this cannot be said
A sin, or shame, or loss of maidenhead,
Yet this enjoys before it woo,
And pamp'rd swells with one blood made two,
And this, alas, is more than we can do.
Oh stay, three lives in one flea spare,
Where we almost, nay more than married are:
This flea is you and I, and this
Our marriage bed, and marriage temple is...*

John Donne, to his lover circa 1600



Mast cell disorders

Mastocytosis (urticaria pigmentosa) There are too many mast cells in the skin (\pm other organs), eg in nests or as a single mastocytoma. These release histamine, leucotrienes, and heparin (this has systemic effects) and proteases (which have local effects). Adults usually express activating mutations of the stem cell factor receptor 'c-kit'. medline:1129852

Skin lesions in adults are typically red-brown macules/papules which urticate if scratched (Darier's sign) and are extremely itchy. Systemic features: headache, palpitations, syncope, flushing, wheeze, diarrhoea, hepatosplenomegaly, osteoporosis, osteosclerosis, anaemia, eosinophilia, and, rarely, mast cell leukaemia. Lancet 2084 & medline:1137768

Diagnosis Skin biopsy; \uparrow urinary histamine, and \uparrow histamine metabolites.

Tests Urinary histamine may be \uparrow (helps if no skin signs). Labs may also report *N*-methylhistamine (MH) and *N*-methylimidazoleacetic acid (MIMA).

Differential diagnoses: Sarcoidosis; histiocytosis.

Treatment Avoid triggers (eg aspirin, heat, cold). H_1 & H_2 antihistamines, disodium cromoglycate. Nd:YAG lasers have been tried. medline:1144609

Lasers in dermatology

Lasers (light **a**mplification by **s**timulated **e**mission of **r**adiation) have an increasing rôle, and it is worth knowing who may benefit from them. The destructive energy of lasers can be concentrated in time and space, and the light's wavelength can be adjusted to match the target lesion. Energy can be selectively taken up by targets as long as the volume delivered is less than the target's capacity to absorb it. If this limit is exceeded, unnecessary destruction is caused. Variables to specify—an example from treating keloid scars: wavelength 585nm, pulse duration 0.45msec, spot size 5mm, mean fluence per pulse 7 Joules/cm², treatment interval: 6 weeks. **NB:** melanin in dark skins may be a problematic competing chromophore.

Photoacoustic damage is produced by shock waves from very intense energy delivered over picoseconds.

Photocoagulation implies less rapid, and less intense treatment.

Selective photothermolysis is somewhere between the above 2 categories. It may produce minimum scarring. Modification of the duration of delivery is possible (Q-switching and mode-locking).

Examples of laser-treatable skin disorders

Port wine stain Best treated when young (smaller, smoother lesions).

Tuberous sclerosis Angiofibromata (misnamed adenoma sebaceum) can cover much of the face.

Strawberry naevi These do *not* need treatment, unless they fail to resolve spontaneously. If they ulcerate, get expert advice.

Flat, pigmented lesions. These are problematic because it is essential to know that they are benign.

Tattoos If multicoloured, multiple wavelengths must be used.

Scars Raised, red, nodular *keloid scars* may appear ugly (and itchy); lasers can help somewhat. *Acne scars* (atrophic or 'ice-pick'): randomized comparisons of CO₂ vs YAG laser are needed. medline:1127973

9. Orthopaedics and trauma

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Relevant pages in other chapters Any medical or surgical illness from acute abdomen to zoster may present to the Emergency Department. So every page here and in *OHCM* has relevance. ▶ See *Pre-hospital care* (p 780–806). Childhood poisoning, p 252–4; (adults, *OHCM* p 824–30).

Internet orthopaedics www.worldortho.com; textbook: www.medmedia.com/med.htm
Images: www.njnet.com/~embbs/xray/xr.html; www.trauma.org/imagebank/imagebank.htm
www.trauma.org/imagebank/imagebank.html **Emergency medicine** www.emedhome.com

▶ Many detailed treatments are described in this chapter, and it is not envisaged that the inexperienced doctor will try them out except under appropriate supervision. The importance of enlisting early expert help (either at once or, if appropriate, by calling the patient back to the next morning's clinic) cannot be overemphasized.

'All these hags'

Once, after a stay in hospital with a fracture, a patient who was not quite as deaf as she was supposed to be, told one of us (JML) how she had overheard a newly arrived orthopaedic surgeon say to the Ward nurse "What are we going to do with all these hags?" As he progressed down the ward, turning first to the left and then to the beds on the right, his mood became morose, then black—as if he was getting angry that all these 'hags' were clogging up his beds, preventing his scientific endeavours. But what was really happening, my patient suspected, was that, as he turned to left and right, he was really nodding goodbye to his humanity, and, dimly aware of this, he was angry to see it go. On this view, these rows of hags were like buoys in the night, marking his passage out of our world. We all make this trip. Is there any way back? The process of becoming a doctor takes us away from the very people we first wanted to serve. Must medicine take the brightest and the best and turn us into quasi monsters?

The answer to these questions came unexpectedly in the months that followed: sheer pressure of work drove this patient's observations out of my mind. It was winter, and there was 'flu. In the unnatural twilight of a snowy day I drifted from bed to bed in a stupor of exhaustion with a deepening sense of a collapse that could not be put off. . . . It was all I could do to climb into bed: but when I awoke, I found I had somehow climbed into a patient's bed, who had kindly moved over to make space for me, and was now looking at me with concern in her eyes. Herein lay the answer: the hag must make room for the doctor, and the doctor must make room for the hag: *we are all in the same bed.*

*A&E nomenclatures

A&E stands for Accident and Emergency. Increasingly, the word accident is being dropped, and **emergency medicine** has emerged as a speciality in its own right—which centres on the 'knowledge and skills required for the prevention, diagnosis, and management of the acute and urgent aspects of illness and injury affecting patients of all age groups with a full spectrum of physical and behavioural disorders. It is a speciality in which time is critical'.—D Fatovich 2002 *BMJ* i 658

The neck

Examination ►► After trauma, the neck must be immobilized, eg with sandbags* and tape, until a lateral cervical spine radiograph has been seen. NB: if very restless, use hard collar only, as otherwise the neck is vulnerable when the body moves on an immobilized head.

The posture of the neck and any bone tenderness are noted. The range of movements: flexion; extension (mainly atlanto-occipital joint); rotation (mainly atlanto-axial joint); lateral flexion (whole of cervical spine). Rotation is the movement most commonly affected. The arms are examined for weakness that might signify root lesions (shoulder abduction C5; elbow flexion C5, 6; elbow extension C6, 7; wrist extension C6, 7; wrist flexion C7, 8; grip and abduction of fingers against resistance T1). Reflexes are examined: biceps C5, 6; supinator C5, 6; triceps C7. Dermatomes are illustrated on p 723. If cord compression is suspected examine the lower limbs for signs of this (see p 608)—eg upgoing plantars, hyperreflexia.

Spasmodic torticollis (Cervical dystonia) The commonest adult focal dystonia. Episodes of sudden-onset of a stiff painful neck with torticollis are due to trapezius and sternocleidomastoid spasm. **Causes:** Genetic; trauma. **Treatment:** If not self-limiting, heat, manipulation, relaxants, and analgesia may help. If severe, wearing a collar may help initial discomfort but may prolong symptoms. Botulinum toxin (p 522) may help for 4 months. Adjuncts: anticholinergics, benzodiazepines, baclofen. medline:957738

Infantile torticollis This may result from birth damage to sternocleidomastoid. **Typical age:** 6–36mths. **Signs:** Tilted head (ear nearer shoulder on affected side). ♂:♀=3:2. There is retarded facial growth on the affected side, hence facial asymmetry. Early, there is a tumour-like thickening in the muscle (may be palpable). **Differential:** Spasmus nutans (head nodding; torticollis; nystagmus); acquired late-infancy torticollis from rare CNS disorders (eg gangliocytoma). If there is an associated muscle mass biopsy may be reassuring by showing a benign fibrous lesion. medline:272516 **Treatment:** Self-limiting in 97%. If persistent, physiotherapy helps by lengthening the muscle; division at its lower end is more drastic. medline:781459

Cervical rib Congenital development of the costal process of the C7 vertebra is often asymptomatic but may cause thoracic outlet compression. Similar symptoms without radiological abnormality is called scalenus or 1st rib syndrome. Thoracic outlet compression involves the lowest trunk of the brachial plexus (p 724) ± the subclavian artery. Pain or numbness may be felt in hand or forearm (often on the ulnar side); there may be hand weakness and muscle wasting (thenar or hypothenar). **Diagnosis:** Weak radial pulse ± forearm cyanosis. Radiographs may not reveal cervical ribs, as symptoms may be caused by fibrous bands. Arteriography may show subclavian compression. **Treatment:** Physiotherapy to strengthen the shoulder elevators may improve symptoms, but rib removal or band division may be needed.

Prolapsed cervical disc Central protrusions (typically of C5,6 & C6,7) may give symptoms of spinal cord compression (p 608; refer to neurosurgeon). Posterolateral protrusions may cause a stiff neck, pain radiating to the arm, weakness of muscles affected by the nerve root, and depressed reflexes. **Tests:** MRI is the preferred image. **Treatment** is with NSAIDs, and a collar. As pain subsides, physiotherapy may help to restore mobility. Surgery is occasionally indicated, in the light of CT/MRI findings

*If there is marked kyphosis, avoid hard collars to stabilize the spine, as the extension produced may be more than is natural for the patient: so collars may cause spinal injury: www.txt.co.uk/ful.cfm?108

Cervical spine images

In major trauma, the first radiograph to do after resuscitation is a cross-table lateral of the cervical spine. All seven cervical vertebrae must be seen, along with the C7–T1 junction: do not accept an incomplete image—oblique views of C7–T1 may be needed. Rarely, subluxations do not show up without flexion and extension views (perform only on the advice of a senior colleague—the patient should move their own muscles; do not rely on passive movements).

- ▶ If a cervical spine injury is suspected you must immobilize the neck until it is excluded.
- ▶ A cross-table lateral in the best hands will miss $\leq 15\%$ of injuries.

When examining the image, follow 4 simple steps:

- 1 Alignment:** Of anterior and posterior vertebral body, posterior spinal canal, and spinous processes. A step of $<25\%$ of a vertebral body implies unifacet dislocation; if $>50\%$ it is bifacetal. 40% of those <7 yrs have anterior displacement of C2 on C3 (pseudosubluxation); still present in 20% up to 16 yrs. 15% show this with C3 on C4. In this physiological subluxation, the posterior spinal line is maintained. Angulation between vertebrae $>10\%$ is abnormal.
- 2 Bone contour:** Trace around each vertebra individually. Look for avulsion fractures of the body or spinous process. A wedge fracture is present if the anterior height differs from posterior by >3 mm. Check odontoid (open mouth view but can be seen on lateral view); don't mistake vertical cleft between incisors for a peg fracture!—epiphyses in children can be mistaken for fractures; the distance between the odontoid and anterior arch of C1 should be <3 mm (may be increased in children). Type 1 fractures are of the odontoid peg, type 2 of its base, and type 3 fractures extend into the body of C2.
- 3 Cartilages:** Check that intervertebral disc space margins are parallel.
- 4 Soft tissues:** If the space between the lower anterior border of C3 and the pharyngeal shadow is >5 mm suspect retropharyngeal swelling (haemorrhage; abscess)—this is often a useful indirect evidence of a C2 fracture. Space between lower cervical vertebrae and trachea should be <1 vertebral body.

Other neck pathologies

Cervical spondylosis (See *OHCM* p 384.)

Cervical spondylolisthesis

This is displacement of one vertebra upon the one below. *Causes:*

- 1** Congenital failure of fusion of odontoid process with the axis or fracture of the odontoid process (skull, atlas, and odontoid process slip forward on axis).
- 2** Inflammation softens the transverse ligament of atlas (eg rheumatoid or complicating throat infections), the atlas slips forward upon the axis.
- 3** Instability after injuries. The most important consequence of spondylolisthesis is the possibility of spinal cord compression. Treatments used include traction, immobilization in plaster jackets and spinal fusion.

The shoulder

Anatomically, the glenohumeral joint is lax, depending on surrounding rotator cuff muscles for stability—ie the sheath of tendons of subscapularis (internal rotation) infraspinatus (external rotation), and teres minor (external rotation + extension). Biceps' long head traverses the cuff, attaching to the top of the glenoid cavity.

History Any trauma? Where is the pain: shoulder? Neck? Does shoulder movement make it worse? (if all movements worsen pain, suspect arthritis or capsulitis; if only some movements, suspect impingement). General health ok? (If aches and pains all over, is it fibromyalgia or polymyalgia?)

Examination Strip to waist. To assess glenohumeral movement, feel the lower half of the scapula to estimate degrees of scapular rotation over the thorax. Half the range of normal abduction is achieved by scapula movement. Test *abduction* by raising hands from the sides sideways to above the head; *flexion* by raising hands forwards and upwards; *extension* by backward movement of elbows; *external rotation* by holding elbows against the sides flexed at 90° and moving the hands outwards (normal 80°); and *internal rotation* by placing the back of the hand against the lumbar spine and moving the elbows forward; an easier test is to assess how far behind her back she can reach: 'imagine you are doing up a bra at the back'.

The muscles used for movement at the shoulder joint

- *Flexion* (forward movement): pectoralis major, deltoid, coracobrachialis.
- *Extension*: deltoid (latissimus dorsi, pectoralis major, and teres major begin the extension if the shoulder starts out flexed).
- *Abduction*: supraspinatus (for first 15°); deltoid thereafter.
- *Adduction*: pectoralis major, latissimus dorsi, teres major, subscapularis.
- *Medial rotation*: pec. major, deltoid, latissimus dorsi, teres major, subscapularis.
- *Lateral rotation*: deltoid, teres minor, infraspinatus.

NB: rotator cuff muscles are most important in rotation: subscapularis, teres minor, infraspinatus (supraspinatus is also part of the rotator cuff).

Scapula movement on the chest wall NB: serratus anterior prevents 'winging' of the scapula as pressure is placed on the outstretched hand.

- *Elevation*: (shrug shoulders) levator scapulae, trapezius
- *Depression*: serratus anterior, pectoralis minor
- *Forward action* (eg punch): serratus anterior; pectoralis major
- *Retraction* (brace shoulders): trapezius, rhomboid.

Recurrent dislocation of the shoulder (For initial dislocation, see 696.) There are 2 types: **Atraumatic**: (5%) The patient is often a teenager with no history of trauma, but having general joint laxity. Remember **BRAV**: bilateral; treat by rehabilitation; atraumatic; dislocates in all directions. **Traumatic**: Dislocation is usually anterior (sometimes inferior, rarely posterior) and secondary to trauma (may be mild). Remember **BUST**: Bankart lesion (see below); unidirectional; surgical treatment; traumatic. Abduction + lateral rotation of the arm (eg donning a coat) may cause dislocation. The capsule is attached to the neck of scapula but detached from the glenoid labrum (Bankart lesion). There may be a posterolateral 'dent' in the humeral head (seen on radiographs with arm medially rotated). Treatment (open or arthroscopic) in Bankart repair (sew capsule back to glenoid) or Putti-Platt operation (the subscapularis tendon is shortened). medline 1095410

With the rarer recurrent posterior dislocation, the capsule is torn from the back of the neck of scapula, the humeral dent is superomedial, and is abduction & medial rotation which causes dislocation (eg seizure).

Treatment: Surgery—if prompt closed reduction fails. medline 1075092
Recurrent *subluxation* is also recognized (disabling and difficult to treat).

The painful shoulder

'There is little evidence to support or refute the efficacy of common interventions for shoulder pain. . .'

meta-analysis
medline: 107964

Remember that the neck may refer pain via C5 to the deltoid region and via C6, C7, and C8 to the superior border of the scapula.

Rotator cuff tears Tears in supraspinatus tendon (or adjacent subscapularis and infraspinatus) may be from degeneration, or, less commonly, to a sudden jolt or fall. Partial tears cause a painful arc (below); complete tears limit shoulder abduction to the 45–60° given by scapular rotation. The arm is passively abducted beyond 90° deltoid's contribution to abduction comes into play, which is then possible from this point. Full-range passive movement is present. Pain is felt at the shoulder-tip and upper arm; there is tenderness under the acromion. *Typical age:* >40yrs. *Tests:* Ultrasound, MRI, or MRI-arthrography reveals communication between joint capsule and subacromial bursa. www.txt.co.uk/full.cfm?1076 *Treatment:* Incomplete: expectant. Complete: prompt repair (eg arthroscopic). medline:110604

Painful arc syndrome This is pain on abduction between 45° and 160°. Causes of pain on abduction:

- 1 **Supraspinatus tendinopathy** or partial rupture of supraspinatus tendon gives pain reproduced by adducting pressure on the partially abducted arm. *Typical age:* 35–60. Only a proportion will have a painful arc (others have increasing pain up to full abduction), which is why the term *impingement syndrome* (as the tendon catches under the acromion during abduction between, eg 70° and 140°) is preferred. *Treatment:* Active shoulder movement with physiotherapy and pain relief; subacromial bursa injection of steroid, eg triamcinolone acetonide 40mg with local anaesthetic; arthroscopic acromioplasty.
- 2 **Calcifying tendinopathy:** *Typical age:* 40. There is acute inflammation of supraspinatus. Pain is maximal during the phase of resorption. *Treatment:* Physiotherapy; NSAIDs; steroid injection; rarely, excision of calcium.
- 3 **Acromioclavicular joint osteoarthritis (AC joint OA):** This is common. *Treatment:* steroid injections or excision of the lateral end of the clavicle.

Tendinopathy of long head of biceps Pain is felt in the anterior shoulder and is characteristically worse on forced contraction of biceps. *Treatment:* Pain relief; hydrocortisone injection to the tendon may give relief but risks tendon rupture. Technique: p 658.

Rupture of long head of biceps Discomfort occurs after 'something has gone' when lifting or pulling. A 'ball' appears in the muscle on elbow flexion. *Treatment:* Repair is rarely indicated as function remains.

612

Frozen shoulder (adhesive capsulitis) may follow modest injury in older people. Pain may be severe and worse at night (eg unable to lie on one side). Active and passive movement range is reduced. Abduction↓ (<90°), external rotation↓ (<30°). It may be associated with cervical spondylosis (more global restriction of movement). *Treatment:* NSAIDs, intra-articular steroid, physiotherapy, manipulation. Meta-analyses of randomized trials have said not to support any one option, but they did not include Der Windt's trial which strongly favoured injections. Resolution may take years. E-BM 1999 4 1

Shoulder osteoarthritis This is not so common as hip or knee OA. Good success rates are being achieved by joint replacement—complications: infection, dislocation, loosening, periprosthetic humeral/glenoid fracture, nerve injury, prosthetic fracture; ectopic ossification. www.txt.co.uk/sum.cfm?1076

The elbow

The normal range of flexion and extension at the elbow is 0–150°. With the elbow flexed, supination and rotation of 90° should be possible. Pain at the elbow and middle outer aspect of the arm may radiate from the shoulder.

Tennis elbow There is inflammation where the common extensor tendon arises from the lateral epicondyle of the humerus (lateral epicondylitis—better named as a teno-periostitis) ± rupture of aponeurotic fibres. This is usually caused by strain. **Presentation:** Pain is worst when the tendon is most stretched (wrist and finger flexion with hand pronated). Pain is felt at the front of the lateral condyle. Ask the patient to extend the wrist, and then pull on the hand to elicit pain. **Radiography:** Nonspecific. **Treatment:** Pain often subsides in time, but some favour injection of the tendon origin (p 658).

medline 10514160 If this fails, physiotherapy may help, or an epicondylitis brace with severe disability, stripping of the common extensor origin from the bone and then allowing it to fall back in place may give relief. Meta-analyses are equivocal.

medline:138817

Golfer's elbow This is medial humeral epicondylitis. It is less common than tennis elbow. Steroid injection may help, but be wary of the ulnar nerve; the brachial artery is also nearby.

medline 1070898

Student's elbow This is a traumatic bursitis following pressure on the elbows, eg while engrossed in a long book. There is pain and swelling behind the olecranon. Other causes are septic and gouty bursitis (look for tophi). The bursa should be aspirated (send fluid for Gram stain and microscopy for crystals). Traumatic bursitis may then be injected with hydrocortisone. Septic bursitis should be formally drained.

Ulnar neuritis Osteoarthritic or rheumatoid narrowing of the ulnar groove and constriction of the ulnar nerve as it passes behind the medial epicondyle, or friction of the ulnar nerve due to cubitus valgus (a possible sequel to childhood supracondylar fractures) can cause fibrosis of the ulnar nerve and ulnar neuropathy. **Presentation:** Sensory symptoms usually occur first, eg ↓ sensation over the little finger and medial half of ring finger. Patients may experience clumsiness of the hand and weakness of the small muscles of the hand innervated by the ulnar nerve (adductor pollicis, interossei, abductor digiti minimi and opponens digiti minimi). **Tests:** Nerve conduction studies will confirm the site of the lesion. **Treatment:** Operative release of the nerve to lie in a new channel in front of the elbow.

Deformities *Cubitus valgus:* The normal degree of valgus ('carrying angle') at the elbow is 10° in ♂, and 15° in ♀. Fractures at the lower end of the humerus or interference with the lateral epiphyseal growth plate may cause the angle to be greater. As a result ulnar neuritis and osteoarthritis may occur. Treat if necessary. Association: Turner's.

Cubitus varus. This may occur after poorly reduced supracondylar fracture.

Osteoarthritis of the elbow Osteochondritis dissecans and fracture involving the joint are risk factors. **Tests:** Flexion and extension are usually impaired but rotation is full. **Treatment:** Operation is rarely indicated, but if pain is felt in the lateral compartment, the head of radius may be excised. Loose bodies may be removed if causing pain or locking of the joint.

The wrist and hand

Dupuytren's contracture Progressive, painless fibrotic thickening of the palmar fascia (aponeurosis) with skin puckering and tethering. **Causes/associations:** Multifactorial, eg genetic (autosomal dominant pattern), alcoholism, antiepileptics, Peyronie's disease, knuckle pads. Ring and little fingers are chiefly affected (\pm plantar fascia). It is often bilateral and symmetrical. As thickening occurs there may be MCP joint flexion. If interphalangeal joints are affected the hand may be quite disabled. Surgery (eg fasciectomy) aims to remove affected palmar fascia and hence prevent progression. As a guide, if he cannot place his palm flat on a flat surface, surgical referral is wise. There is tendency for recurrence. Try to avoid finger amputations. medline:1099180

Ganglia These smooth, multilocular jelly-like swellings are bulges of synovium, eg at the wrist. They communicate with joint capsules or tendon sheaths. **Treatment** is not needed unless they cause pain or pressure (eg on median or ulnar nerve at wrist, or lateral popliteal nerve at the knee). They may disappear spontaneously. Local pressure may disperse them (traditionally a blow from a Bible). Aspiration via a wide-bore needle may work, or they may be dissected out; problems include painful scars, neurovascular damage (esp. in volar wrist ganglia), and recurrence. medline:1110348

De Quervain's syndrome (Stenosing tenovaginitis) There is pain over the styloid process of the radius and thickening of the tendons of abductor pollicis longus and extensor pollicis brevis. Pain is worst when these tendons are stretched (eg lifting a teapot) and is elicited by forced adduction and flexion of the thumb into the palm. (Finckelstein's sign). Pain is more proximal than that from osteoarthritis of the first carpometacarpal joint. **Cause:** unknown but it commonly follows activities involving much use of the tendons (eg wringing clothes). **Treatment:** First try NSAIDs or hydrocortisone injection around the tendons in their sheaths. If injection (p 658) and rest fail to relieve, decompression of the tendons is provided by deroofing or slitting the tendon sheaths. >80% do well post-op. medline 11386780 & 877654

Trigger finger (Tendon nodules) Disproportion of the tendon to its sheath giving a fixed flexion deformity. Ring and middle fingers are most commonly affected, and the thumb especially in babies and children. Full extension of the affected digit cannot be achieved by the muscles of the hand, and if aided by the other hand, a 'click' may be felt in the flexor tendon. As extension occurs, the nodule moves with the flexor tendon, but then becomes jammed on the proximal side of the pulley, and has to be flicked straight, so producing triggering. Nodules are associated with rheumatoid arthritis. **Treatment:** Unless the patient is a child, or has renal failure or diabetes, steroid injection into the region of the nodule, under local pressure, but not into the nodule itself (eg 1mL of Depomedrone[®] + lidocaine) may be tried. Risk of recurrence is high, so surgery may be needed (eg as an 'office procedure' to release the 'A1' pulley). medline:9562759 & 114765

Volkmann's ischaemic contracture Follows interruption of the brachial artery near the elbow (eg after supracondylar fracture of humerus, p 698). Muscle necrosis (especially flexor pollicis longus and flexor digitorum profundus) results in contraction and fibrosis causing a flexion deformity at wrist and elbow, with forearm pronation, wrist flexion, thumb flexion & adduction, digital metacarpophalangeal joint extension, and interphalangeal joint flexion. Suspect if a damaged arm is blue, with no radial pulse, and passive finger extension is painful (this is the crucial sign). **Treatment:** Remove constricting splint, warm other limbs (promote vasodilatation). If pulse doesn't return within 30min, explore the artery. **Treating contractures:** release of compressed nerves \pm tendon lengthening and transfers to restore lost function. medline:97424

Examination of the back

History The commonest orthopaedic complaint is back pain. Attention should be paid to the nature of the pain, exacerbating and relieving factors and the history of onset. It is important to know if bowel or bladder function is affected (this signifies cord or cauda equina compression and should set your alarm bells ringing loudly: refer at once). With low back pains it is not uncommon for pain to radiate down the leg (sciatica): such pain may be accompanied by root signs (see below).

Examination With the patient standing and wearing only bra & pants inspect the back for abnormality and palpate for local tenderness and deformity. Movements assessed are *forward flexion* (stretch forward to touch toes with knees straight—look to see how much movement is due to back flexion and how much by flexion at the hips—with back flexion the back has a gently rounded contour. Look for a rib lump, a sign of scoliosis); *extension* (arch spine backwards); *lateral flexion* (lean sideways so hand moves down corresponding thigh) and *rotation* (keep pelvis fixed but move shoulders round to each side in turn). Movement at the costovertebral joints is assessed by the difference in chest expansion between maximal inspiration and expiration (normal: 5cm). Iliac crests are grasped by the examiner and compressed to move sacroiliac joints and see if this reproduces the pain.

Note: On bending fully forwards, expansion of a line drawn from 10cm above L5 to 5cm below it by <5cm is firm and quantifiable evidence of movement restriction.

Lower limbs are compared for leg length discrepancy and muscle wasting (measure thigh and calf circumference), for weakness, sensory loss, and normal reflexes (knee jerk is mainly L4, ankle jerk S1, plantars should be downgoing).

Straight leg raising *Why?* To test for a mechanical cause of sciatic pain. *How?* Keeping the knee extended, lift the patient's leg off the couch and note the angle to which the leg can be raised before eliciting pain. If <45° Lasègue's sign is said to be positive. *Mechanism?* This stretches the sciatic nerve and causes root pain (a characteristic lancinating pain distributed in the relevant dermatome, and made worse by coughing or sneezing).

Alternatively The method described above often elicits pain in the hypersensitive. Alternatively sit the patient up in bed with legs out in front. This can be done while examining another system, and may prove pain-free.

Other parts of the body examined These are the iliac fossae (important in days when tuberculous psoas abscesses were common), abdomen, pelvis and rectum, and major arteries. The commonest tumours to metastasize to bone are: breast, bronchus, kidney, thyroid, and prostate so it may be relevant to examine these.

Tests In acute low back pain, tests are not usually needed in the first 2 weeks. *First line.* Hb, ESR (if high, think of metastases or myeloma, and do electrophoresis ± bone marrow), alkaline phosphatase (high in Paget's disease and tumours), calcium. Radiographs: AP, lateral of lumbar spine and pelvis—but note: the dose of radiation is *not* negligible: use x-rays sparingly. *Second line:* MRI (good as non-invasive, and high quality images) enables the cauda equina to be seen (look for disc protrusion, tumours, or spinal stenosis). Ultrasound and CT can illustrate the spinal canal. Radioisotope scans may reveal 'hot spots' of tumour or pyogenic infection.

Kyphosis and scoliosis

Kyphosis is flexion deformity of the spine. The cervical spine is usually held slightly extended (lordosis), the thoracic spine flexed, and lumbar spine again extended. Conditions affecting the architecture of the spine may be a cause of kyphosis, eg developmental deficiency of vertebra, Scheuermann's or Calvé's osteochondritis (p 650 & p 652), wedge fractures of vertebrae (osteoporosis, metastatic carcinoma, TB), ankylosing spondylitis. Treat the underlying disorder if indicated. NB: congenital kyphosis is much less common than congenital scoliosis, but is more serious, as cord compression and paraplegia sometimes develop rapidly, eg during adolescence. *medicine* 105:355-87

Scoliosis This is lateral spinal curvature + vertebral and rib rotation. The commonest cause is muscle spasm (eg with sciatica). Classification:

- 1 Idiopathic (this is seen both in infants and adolescents).
- 2 Congenital (due to bony anomalies).
- 3 Neuromuscular (eg postural, polio, syringomyelia, torsion dystonia). Unequal muscle spasm or paralysis exerts unequal pull on the spine (curve concave to the stronger side).
- 4 Rarities: spinal tumours, spondylolisthesis, arthrogryphosis, enchondromatosis, osteogenesis imperfecta (p 648), neurofibromatosis (*OHCM* p 390), Chiari malformation (*OHCM* p 720), Duchenne muscular dystrophy, Friedreich's ataxia, Marfan's syndrome (*OHCM* p 730), Pompe's disease (p 756).

Idiopathic scoliosis There is lateral curvature of the thoracic or lumbar spine $>10^\circ$. Adolescent idiopathic scoliosis (AIS) is the most common spinal deformity; girls are more often affected than boys—and they are often taller than their peers at presentation (but not ultimately). Complications in later life revolve around pain, unsightliness, and impaired lung function. Thoracic problems tend to give more severe deformity than lumbar. Rib deformity causes a characteristic hump on the convex side of the curve which becomes manifest on asking the patient to bend forwards. Curvature increases while the affected person continues to grow, so usually the earlier the onset the worse the deformity. Since the advent of screening, scoliosis has been detected in 1.5–3%; of these only ~6% progress: the younger the child, the more the risk of progression—and double curves progress more than single curves. Thoracolumbar or lumbar curves progress the least. **Treatment** is needed in 2.75/1000 screened. Where curvatures are $<45\text{--}50^\circ$ and growth is completed, or $<29^\circ$ if growth is in progress, the situation is monitored regularly: eg every 3 months during growth, and every 8 months thereafter.¹ When curvatures are progressing, attempts to halt it may be made using a Boston or Milwaukee distraction-derotation brace (particularly for double curves, or if the apex of the curve is higher than the 8th thoracic vertebra¹) until the child is old enough for any spinal surgery that may be indicated (eg Luque rods and wires) or Cottrill–Dubousset instrumentation (Harrington's metal rods are now rarely used, but paralysis, the most feared complication of spinal surgery occurs in only 0.23%, compared with ~0.7% with other operations). NB: it can be hard to persuade a child to wear a brace for the $\geq 20\text{h/day}$ that is optimal!—but bracing *does* work. There is no effective treatment for established deformity in adults.

► When scoliosis in youth gives pain (especially at night), exclude osteoid osteoma, osteoblastoma, spondylolisthesis, and spinal tumours.

¹ Boston braces worn for $>18\text{h/day}$ do prevent progression of large curves at a mean of 9.8 years after bracing is discontinued: *J Wiley* 2000 *Spine* 25 2326 [10984784]

Causes of back pain

Sinister back pain: ► p 626

Backache is usually from low back strain or degenerative disease. *Other causes:* retroperitoneal disease (duodenal ulcer, pancreas cancer, aneurysm; pain is often lumbodorsal and spine movements pain-free & full). Other causes: spine neoplasia; infection; disc lesions, arthritis, osteoporosis, osteomalacia.

Mechanical pain The spine is a complex series of articulations, with spongy discs between vertebral bodies acting as shock absorbers, and a multitude of articulating facet joints. Problems in one part affect function of the whole, and spasm of vertebral muscles can cause much pain. Upright posture provokes big forces on the spine, eg when lifting and discs may rupture (if young), vertebrae fracture (elderly) or soft tissues tear (low back strain). NB: with low back strain the exact mechanism may be elusive; if there are no sinister features (p 624) take time to explain the diagnosis and management (p 624); x-ray if unimproved eg at 6 weeks.

Disc prolapse Lumbar discs are those most likely to rupture (esp. the lowest 2). Typically, one is seized by severe pain on coughing, sneezing, or twisting a few days after back strain (onset may be insidious). Pain may be confined to the lower lumbar area (lumbago), or may radiate to buttock or leg (sciatica) if the herniated nucleus pulposus compresses a nerve root.

Signs: Forward flexion (p 618) and extension limited, \pm lateral flexion—unilaterally and inconstantly. With L5–S1 prolapse, S1 root compression causes calf pain, weak foot plantar flexion, \downarrow sensation (pinprick) over sole of foot and back of calf, and \downarrow ankle jerk. With L4–5 prolapse, hallux extension is weak and sensation \downarrow on outer dorsum of foot. If lower lumbar discs prolapse centrally, cauda equina compression (p 728) may occur. Danger signs: •Saddle anaesthesia •Incontinence/retention •Paralysis.

Tests: MRI (or CT or myelography) if intervention is contemplated, eg cauda equina compression, or if rest fails and symptoms are severe, with CNS signs such as reflex or sensory changes, or muscle wasting.

Treatment of disc prolapse: Brief rest with early mobilization + pain relief (p 624) is all that is needed in $\sim 90\%$ (\pm physiotherapy). Discectomy is indicated with cauda equina syndrome, progressive muscular weakness and continuing pain.

40 trials
medline:1048851

Spondylolisthesis There is displacement (usually forward) of one lumbar vertebra upon the one below. *Causes:* Spondylolysis (defect in pars interarticularis of neural arch); congenital malformation of articular processes osteoarthritis of posterior facet joints (older folk). Onset of pain with or without sciatica is often in adolescence. *Diagnosis* is by plain radiographs.

Treatment may involve wearing a corset, or nerve release and spinal fusion.

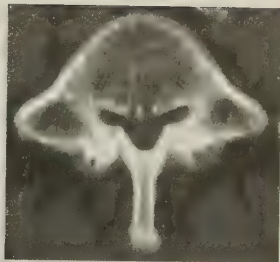
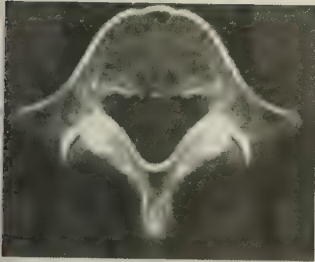
Lumbar spinal stenosis (LSS) and lateral recess stenosis

Facet joint osteoarthritis (the only synovial joints in the back) may produce generalized narrowing of the lumbar spinal canal or simply of its lateral recesses. Unlike the pain of lumbar disc prolapse, this causes:

- Pain worse on walking with aching and heaviness in one or both legs causing the person to stop walking ('spinal claudication').
- Pain on extension. •Negative straight leg raising test. •Few CNS signs.

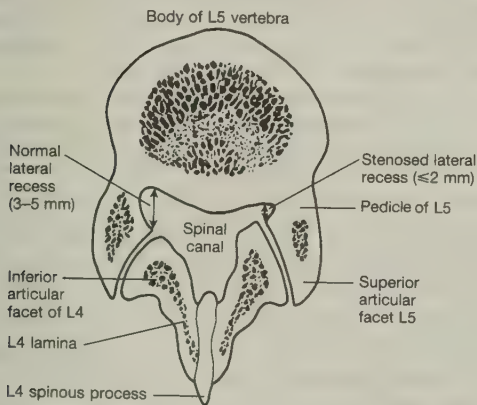
Tests: MRI (CT and myelography are less good options).

Treatment: Canal decompression (removing its posterior wall) gives good results if NSAIDs, epidural steroid injections, and corsets (to prevent exaggerating the lumbar lordosis of standing) fail to help.



Cross-sectional CT images at the L5 level, one with spinal stenosis. Observe the difference in spinal canal shape and dimensions as a result of osteoarthritis of the facet joints posterolaterally. Other factors that may contribute to acquired spinal stenosis include disc prolapse, spondylolisthesis, and congenitally short pedicles. Identify the spinal canal contents at this level, including the thecal sac and the two L5 nerve root sheaths that occupy the lateral recesses. Anatomy is as outlined below.

Cross-section of vertebral canal at L5 showing lateral recess stenosis



Management of low back pain

Back pain is common. 80% of the Western population suffer it at some stage in their lives. 50/1000 industrial workers have time off work because of it each year. In the UK it accounts for 11 million lost working days/yr (£3.8 billion/yr in lost production). A GP can expect 20 people per 1000 on her list to consult her with it each year, of whom 10–50% are referred to hospital. Fewer than 10% of those referred to a specialist will have surgery. Physiotherapy may be carried out in hospital or general practice.

Most back pain is self-limiting: of those attending GPs, 70% are better after 3 weeks, 90% by 6 weeks, irrespective of treatment. But remember: the cause of back pain can be sinister—malignant disease, local infection, or cord or cauda equina compression—and an approach is needed that detects these promptly. Age is important: in one study only 3% of patients aged between 20 and 55 had 'spinal pathology' (tumour, infection, inflammatory disease) as compared with 11% of those <20yrs, and 19% >55yrs.

Features which may indicate sinister back pain (Causes: p 626)

- Young (<20y) or old (>55)
- Progressive, continuous, non-mechanical pain
- Violent trauma
- Systemically unwell
- Alternating sciatica (both legs involved)
- Drug abuse or HIV +ve
- Bilateral sciatica
- Spine movement in all directions painful
- Weak legs
- Localized bony tenderness
- Weight loss
- CNS deficit at more than one root level
- ESR ↑ (>25mm/h)
- Pain or tenderness of thoracic spine
- Bilateral signs of nerve root tension
- Taking oral steroids
- Past history of neoplasia

► Pain brought on by activity and relieved by rest is *rarely* sinister.

Those with any suggestion of cord or cauda equina compression (p 728), or with deteriorating unilateral signs must be referred at once. Those in whom malignancy or infection is suspected should be referred promptly.

Treating biomechanical back pain 'Get on with your life within the limits of the pain' gives better results than physiotherapy with lateral bending exercises. Avoid bed-rest after the 1st 48h (a board under the mattress helps). Avoid slouching. Advise on avoiding back stress: how to rise from lying, avoid stooping, bending, lifting, and low chairs.

Give good analgesia to break the pain–muscle spasm cycle (paracetamol, up to 4g/24h po, ± NSAID, eg naproxen 250mg/8h po pc—but opioids may be needed at first). Warmth helps, as does swimming in a warm pool. If spasm persists, try diazepam 2mg/8h po for a short while. Antidepressants can help refractory pain, but not level of functioning.

Physiotherapy in the acute phase can ↓ pain and spasm. In convalescence, give education on lifting and exercises to strengthen the back muscles.

Many consult an osteopath or chiropractor for manipulation (as done by some physiotherapists). Studies show manipulation can produce relief of symptoms which is dramatic but may not be longlasting.

If pain does not improve after 2 weeks, do blood tests, and, if abnormal, consider radiographic examination. Later, referral to rehabilitation services, epidural anaesthesia, and corsets may help. Orthopaedic referral may be needed if pain does not resolve with time, for help with diagnosis, for additional treatment, or for reassurance.

Why do some people get intractable back pain?

39% of adults have (or have had) an intermittent or chronic back problem—and much energy is expended on frequently fruitless searches for the causes listed opposite. Radiographs and MRI may be non-specific (sub-clinical disc protrusion, facet joint arthritis, or minor spondylosis). Risk factors for chronicity are:

- Smoking
- Psychological morbidity
- Low income/social class
- Co-existing cardio-respiratory disease
- Poor work conditions
- Number of children (for women *and* men)

Be cautious of blaming sitting at work for chronic back pain. Meta-analyses do not find in favour of this popular association. meta-analysis
medline:11851181

Vascular problems may underlie chronicity (hence associations with smoking and heart disease), and pain may be maintained by involvement of the sympathetic chain, which is thought to mediate hyperaesthesia, hyperpathia (excess pain from minor noxious stimuli), allodynia (pain generated by minor skin stimulation)—but surgical sympathectomy often only provides temporary relief. This implies central neuromodulation of stimuli producing a complex regional pain syndrome. Dorsal horn receptor fields may expand and have their thresholds changed by peripheral injury, so pain is more intense, and appreciated over a wider area than simple anatomy would predict. <http://bmj.com/cgi/content/full/314/7095/1639>

Specific and sometimes sinister back pain

Typical causes 15–30yrs: Prolapsed disc, trauma, fractures, ankylosing spondylitis (p 674), spondylolisthesis (L5 shifts forward on S1), pregnancy.
>30yrs: prolapsed disc, cancer (lung, breast, prostate, thyroid, kidney).
>50yrs: Degenerative, osteoporosis, Paget's, malignancy, myeloma. Lumbar artery atheroma (which may itself cause disc degeneration).

Rarer causes: Spinal stenosis (bony encroachment); cauda equina tumours; spinal infection (usually staphylococcal, also *Proteus*, *E coli*, *Salmonella typhi*, TB). Often no systemic signs of infection.

Spinal tumours These may be of spinal cord, meninges, nerves, or bone. They may be primary, secondary, lymphoma, or myeloma. They may compress the cord, causing pain, lower motor neurone signs at the level of lesion, upper motor neurone signs and sensory loss below; bowel and bladder dysfunction. Peripheral nerve function may be impaired resulting in pain along the course of the nerve, weakness, hyporeflexia & ↓sensation (p 722). With cauda equina involvement there is often urinary retention ± saddle anaesthesia. When the deposit is in the spinal canal and if there is no bone involvement, there may be no pain, just long tract signs. When bones of the back are involved there is progressive constant pain and local destruction of bone. Metastases tend to affect cancellous bone, but focal lesions cannot be seen on radiographs until 50% of bone mass is lost. There may be much muscle spasm and local tenderness to percussion. Bone collapse may result in deformity, or cause cord or nerve compression. **Tests:** Plain radiographs; CT; MRI; isotope bone scans; bone biopsy.

In those with past cancer and current back pain, it is best to do a bone scan first, with plain radiographs of any hot spots suggesting metastases.

Pyogenic spine infections This is a notoriously difficult diagnosis as all signs of infection may be absent (eg no fever, tenderness, or WCC↑), although the ESR is often raised. It may be secondary to other septic foci. Pain occurs, and movement is restricted by spasm. It is usually an infection of the disc space (discitis). **Risk factors:** Diabetes mellitus, immunosuppression, urinary surgery, or catheterization. Half the infections are staphylococcal; *Proteus*, *E coli*, *Salmonella typhi* and TB also occur. **Tests:** Radiographs shows bone rarefaction or erosion, joint space narrowing ± subligamentous new bone formation. Technetium bone scans and MRI are better. **Treatment:** As for osteomyelitis elsewhere (p 644), resting the back with bed rest, brace, or plaster jacket. Surgery may be needed.

Spinal TB Rare in those born in the West, this tends to affect young adults, giving pain, and stiffness of all back movements. ESR↑. Abscesses and cord compression may occur (Pott's paraplegia). Radiographs show narrow disc spaces and local osteoporosis early, with bone destruction leading to wedging of vertebrae later. In the thoracic spine, paraspinal abscesses may be seen on radiographs, and kyphosis on examination; with lower thoracic or lumbar involvement abscess formation may be related to psoas muscle in the flank, or in the iliac fossa. MRI (not CT) is the ideal way to delineate cord compression. Treatment is anti-TB drugs (eg for 1yr; p 278) with abscess drainage. Fixation + bone grafting, or metal rods, plates, or wires needs highly specialized services. If a syrinx (tubular cavity) has developed, complete neural recovery is not expected.

medline:10878776

Central disc protrusion This neurosurgical emergency is suggested by bilateral sciatica, perineal or 'saddle' anaesthesia and disturbance of gut or bladder function. ▶ Prompt decompression may prevent leg paralysis.

Diagnostic triage: which group encompasses your patient?

<i>Simple lumbosacral pain</i>	<i>Signs of nerve root pain</i>	<i>Possibly sinister features</i>
Aged 20–55 with 'mechanical' backache, eg caused by twisting and worse on moving	One leg hurting more than the back	Pain unrelated to mechanical events
Patient well-in-himself: no fever	Radiation to foot/toes	Thoracic pain ± deformity
no weight loss	Hip flexion reproduces the pain in the leg	Past history, eg HIV; TB neoplasia, or steroid use
	Localized neurological signs	Widespread CNS signs
		Weight loss; fever; unwell; no pain, but long tract signs

Signs demanding immediate (same-day) referral: gait or sphincter disturbance; saddle anaesthesia—these suggest cauda equina compression.

For evidence on managing these categories, see www.rcgp.org.uk/backpain/rcbp0019.htm

Hip history and examination

(For hip prostheses, see p 654.)

The movements examined at the hip are flexion (grasp iliac crest to eliminate pelvic rotation), normal is 120° ; abduction—normal is $30\text{--}40^\circ$ (keep hand between anterior superior iliac spines to prevent pelvic tilt); abduction in flexion (normally 70°) and adduction (normally 30° , tested by moving one foot across the other). Normal external and internal rotation is 30° . Internal rotation is often the first movement to be restricted by hip disease.

Questions Are activities of daily living affected?—walking distance, ability to climb stairs (only one at a time possible?), difficulty getting out of low chairs. Remember that pain in the knee may be referred from the hip.

Measurements Apparent leg length disparity (with the lower limbs parallel and in line with the trunk) is called either 'apparent shortening' (eg due to pelvic tilt or fixed adduction deformity—which gives the apparent shortening on that side) or 'apparent lengthening' (eg due to fixed hip abduction). In these cases, there is no true disparity, as detected by measuring between the anterior superior iliac spine and medial malleolus on each side with the pelvis held square and the lower limbs held equally adducted or abducted.

Fixed deformity Joint or muscle contractures prevent limbs from being put in the neutral position. With fixed adduction deformity, the angle between the limb and the transverse axis of the pelvis (line between both anterior superior iliac spines) is $<90^\circ$ but with fixed abduction deformity it is $>90^\circ$. Fixed flexion deformity is detected by **Thomas' test**: Feel for lumbar lordosis* under the side on which fixed flexion deformity is suspected. If none is present, there is no flexion deformity (provided both legs are resting flat on the table). If lordosis is present, flex the sound side maximally (knee to chest). At the end point of flexion, you will feel the lumbar spine press against your hand as the lumbar lordosis straightens out. If there is a fixed flexion deformity, the leg opposite the one being flexed will rise a few degrees off the table, as it is being pulled up as the pelvis rotates. Push down on the thigh of the side being tested to help check whether or not it has been pulled off the table. The angle through which the thigh is raised is the angle of fixed flexion. (NB: to assess the full range of extension, have the patient assume the prone position on the table and then extend the hip.)

www.txt.co.uk/full.cfm?1078

The Trendelenburg test This tests the stability of the hip and the ability to support the pelvis when standing on one leg. In this state, it is normal for the pelvis to rise on the side of the lifted leg. A +ve test is when the pelvis falls on the side of the lifted leg. **Causes.** 1 Abductor muscle paralysis (gluteus medius and minimus); 2 Upward displacement of the greater trochanter (severe coxa vara or dislocated hip); 3 Absence of a stable fulcrum (eg ununited fractures of the neck of the femur).

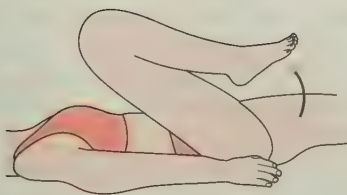
Gait Those with unstable or painful hips use a stick on the opposite side from affected hip. (The reverse is true for knees.)

Other joints to examine Spine, knee, sacroiliac joints/pelvis.

Coxa vara This is the term used to describe a hip in which the angle between the neck and the shaft of femur is less than the normal 125° .

Causes Congenital; slipped upper femoral epiphysis; fracture (trochanteric with malunion, ununited fractures of neck of femur); due to softening of bones (rickets, osteomalacia, Paget's disease). **Consequences.** True shortening of limb; Trendelenburg 'dip' on walking makes the affected person limp.

*Lordosis is lumbar spine anterior convexity (to give a 'hollow back')—when viewed from the side.



Painful hips in children

- ▶ When a child complains of pain in the knee, examine the hip.
- ▶ Is $T^{\circ}\uparrow$? If so, do urgent blood culture & ultrasound-guided aspiration.
- ▶ Think of slipped femoral epiphysis in adolescents.

When a child presents with an unexplained painful limp, examine the hip clinically and radiologically. Admission for observation and rest is usual (\pm traction), and tests are carried out to exclude TB and Perthes' disease. When there has been *unilateral limitation of all hip movements, spontaneous recovery after bed rest, and normal radiology*, a retrospective diagnosis of transient synovitis of the hip (TSOH) is made (known as the 'irritable hip'—the commonest cause of childhood hip pain).^{*} If other joints are involved, consider the diagnosis of juvenile idiopathic arthritis (p 758).

Ultrasound is very helpful in assessment. It can guide needle aspiration when pus is suspected—and may obviate the need for arthrotomy.

Perthes' disease This is osteochondritis of the femoral head and affects those aged 3–11yrs (commonest 4–7yrs). It is bilateral in 10%. $\sigma/\text{f} \approx 4:1$. It presents with pain in hip or knee and causes a limp. On examination all movements at the hip are limited. Early radiographs \pm MRI show widening of the joint space. Later there is a decrease in size of the femoral head with patchy density. Later still, there may be collapse and deformity of the femoral head with new bone formation. Severe deformity of the femoral head risks early arthritis. The younger the patient the better the prognosis. For those with less severe disease ($< \frac{1}{2}$ the femoral head affected on lateral radiographs, and joint space depth well preserved) treatment is bed rest until pain-free, followed by radiographic surveillance. If prognosis poorer ($> \frac{1}{2}$ femoral head affected, narrowing of total joint space) surgery may be indicated.

medline:9393917

Slipped upper femoral epiphysis This affects those aged 10–16yrs. 20% are bilateral. $\sigma/\text{f} \approx 3:1$. 50% of patients are obese. There is displacement through the growth plate with the epiphysis always slipping down and back. It usually presents with limping and pain in the groin, anterior thigh, or knee. Symptoms may be chronic or acute (the latter may represent a very unstable slip, and suggests early fixation may be needed).

On examination flexion, abduction, and medial rotation are limited (eg lying with foot externally rotated). Diagnosis is by a lateral radiograph (may look normal on AP). If untreated, consequences may be avascular necrosis of femoral head or malunion predisposing to arthritis. Minor epiphyseal slips are carefully fixed *in situ*. The rest may undergo a gentle manipulation. Only hips with persisting deformity need femoral osteotomy. medline 3584443

▶ Symptoms may be mild: have a high index of suspicion if in correct age group.

Tubercular arthritis This is rare in the UK, but world-wide there is a resurgence of TB. Children aged 2–5yrs and the elderly are most commonly affected. The symptoms are pain and a limp. All hip movements cause pain and muscle spasm. **Tests:** Early radiographic sign: rarefaction of bone. Subsequently there is fuzziness of joint margins and narrowing of the joint space. Later bony erosion may be seen. Ask about contacts. Check ESR, CXR, and Mantoux test (OHCM p 568). Synovial membrane biopsy confirms the diagnosis of TB arthritis. **Treatment** (eg of hip): rest + anti-TB drugs (p 278), given by experienced personnel. Arthrodesis may be needed if much joint destruction has occurred.

^{*}Protocols exist for assessment of TSOH: occasionally a Perthes' patient slips through: (@10505915)

Developmental dysplasia of the hip (eg hip dislocation)

1.3% of neonates have unstable hips/subluxation. A hip may be normal at birth, and become abnormal later. Incidence (UK): 2 per 1000 live births. ♀/♂ ≈ 6:1; left hip/right hip incidence ≈ 4:1 (bilateral in 1/3). **At-risk babies:**

- Breech birth • Caesar for breech • Other malformations • +ve family history
- Birth wt ↑ • Oligohydramnios • Primip/older mother • Postmaturity

Diagnosis Examine hips of all babies in the 1st days of life and at 6 weeks. With well-trained, well-supported staff this prevents later dysplasia. ^{A. H. J. F. 2002 BMJ 1 103}

► Be alert to CDH throughout childhood surveillance (p 208).

Click test of Ortolani: With the baby supine and relaxed, flex the hips to 90° and knees fully. Place your middle finger over the greater trochanter, and thumb on inner thigh opposite the lesser trochanter. Diagnose a dislocated hip if slow hip abduction produces a palpable (often audible) jerk or jolt (ie more than a click)—as the femoral head slips back into the acetabulum.

The Barlow Manoeuvre: With the pelvis stabilized by one hand, abduct each hip in turn by 45°. Forward pressure by the middle finger causes the femoral head to slip into the acetabulum if the hip is dislocated. If the femoral head slips over the posterior lip of acetabulum and straight back again when pressure is exerted by the thumb it is 'unstable' (ie dislocatable not dislocated). Use both tests but avoid repetitions (may induce instability/dislocation). NB: both tests are problematic, missing up to 2/3 of those later needing surgery.

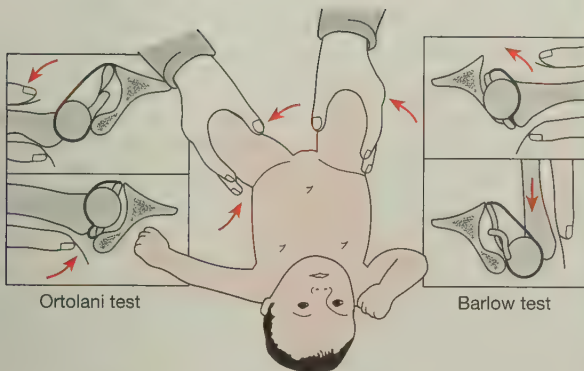
In older children signs may be: delay in walking, abnormal waddling gait (affected leg is shorter), asymmetric thigh creases (extra crease on the affected side), and inability to fully abduct the affected hip. With bilateral involvement the perineum appears wide and lumbar lordosis is increased.

Ultrasound is the image of choice, as it is non-invasive and dynamic. It is unclear if it should be part of general screening. In one series of 4717 babies, 17 needed treatment, and in 5 of these clinical examination missed the diagnosis. 81 had ultrasound abnormalities which did not require treatment. Screening might be confined to high-risk groups (above).

Treatment If neonatal examination suggests instability, use double nappies; reassess at 3 weeks. If still a problem, splint the hips in moderate abduction for 3 months (eg von Rosen splint). Excess abduction may cause avascular necrosis of the head of femur. From 6–18 months closed reduction may be tried using a frame or gallows and traction to gradually abduct the hips to 80°. If this produces full reduction, the legs are then splinted, medially rotated and abducted; if it does not, various operations may be used before 6yrs—aiming to reduce excessive anteversion of the femoral neck (eg rotation osteotomy) and improve the acetabulum so the femoral head lies more deeply in it. From 7–10yrs similar operations may be appropriate (eg for high-lying femoral heads in false sockets). From 11yrs, operations (eg pelvic osteotomy) are principally for pain. Hip replacement may be needed in early adult life for painful osteoarthritis. Arthrodesis is an alternative.

Club foot

Neonatal club foot (talipes equinovarus; ♂/♀ > 1). The foot deformity consists of: **1** Inversion; **2** Adduction of forefoot relative to hindfoot; **3** Equinus (plantarflexion). The foot cannot be passively everted and dorsiflexed through the normal range. Treatment begins within 1 week of birth with weekly foot manipulations, holding it strapped or splinted in position between manipulations. The knee is flexed 90° in a splint to stop the baby drawing up his foot. If treatment has not corrected the foot by ~3 months, operative reduction is carried out. If this does not work, operations on soft tissues and/or bones of the foot may be carried out later (from 2 yrs).



Knee history and examination

► *Pain in the knee may be referred from the hip.* So examination must include the hip. Does internal hip rotation hurt?—so revealing hip pathology.

Ask about trauma, pain, swelling, mobility, locking, clicking, and giving way (eg on exercise—a feature of cruciate ligament injury).

Examine supine with legs fully exposed. Examine for swelling (*Causes:* Bone thickening, fluid, synovial thickening—this feels 'rubbery'). Look for wasting of the quadriceps. The presence of fluid can be confirmed by placing the palm of one hand above the patella over the suprapatellar pouch and thumb and forefinger of the other hand below the patella. Fluid can be moved between the two (hydraulic pressure will be felt) by alternating the source of pressure. If 30–40mL fluid are present it may be possible to feel a 'patellar tap' (ballot patella against neighbouring bones). Patellar taps are absent with small or tense effusions (which may be up to 120mL).

Flexion and extension at the knee vary between persons. Flexion should be sufficient for the heel to touch the buttock. Compare extension with the 'good' side. Medial and lateral ligaments are examined with the knee flexed 20–30° (to relax the posterior capsule), as one hand lifts the ankle off the couch, the other holds the knee just slightly flexed. The knee is stressed in abduction by abducting the ankle with one hand while pushing the knee medially with the hand behind the knee (tests medial ligament). Reverse the pressures to give adducting force to test lateral ligament. If these ligaments are torn the knee joint opens more widely when the relevant ligament is tested (compare knees against each other).

Test the cruciate ligaments with the knee 90° flexed, with the foot placed on couch with you sitting on it (anchor tibia). Place your fingers interlocking behind the knee clasping the sides of leg between the thumbs (each tip on a femoral condyle). With quadriceps relaxed, assess antero-posterior glide of tibia on femur (normal ~0.5cm). The anterior cruciate ligament prevents anterior glide; the posterior prevents posterior glide. Excessive glide in one direction (compare knees) suggests damage to the relevant ligament. Examination should also be performed in 20° of flexion (Lachman's test). A more sensitive test to determine if symptoms are really due to cruciate ligament damage (can be asymptomatic) is the 'pivot shift test': flex the knee, then put it in valgus; now extend it. If the anterior cruciate is ruptured, the knee jumps smartly forwards. (Often hard to elicit, unless very relaxed or under general anaesthesia.)

McMurray's rotation test is an unreliable way of detecting pedunculated tears of menisci. The knee is flexed, the tibia laterally rotated on the femur, then the knee is extended with tibia kept rotated. This is repeated with varying degrees of knee flexion, and then again with the tibia medially rotated on the femur. This manoeuvre is designed to jam the free end of a pedunculated meniscus in the joint—a click being felt and heard and pain experienced by the patient as the jammed tag is released as the knee is straightened. This test may not detect bucket-handle tears (p 710). Note: normal knees commonly produce patellar clicks. Simply eliciting joint-line tenderness may be a more valid test when combined with a history of mechanical locking.

Arthroscopy Arthroscopes enable internal structures of the knee to be seen and a definite diagnosis may be made. They also enable a wide range of operations to be done as day-case surgery.

Pain in the knee

The common symptoms are anterior knee pain or pain and swelling. Anterior knee pain can be due to many causes (see below).

Chondromalacia patellae ('Idiopathic adolescent anterior knee pain syndrome') It particularly affects young women and servicemen. Patella aching is felt after prolonged sitting. Medial retropatellar tenderness and pain on patellofemoral compression occur. **Diagnosis** is clinical but if arthroscopy is performed softening and/or fibrillation of patellar articular cartilage is seen. **MRI** has a rôle.

medline:895861

Treat by vastus medialis strengthening exercises (lie on back with foot externally rotated; lift heel 10cm off the floor \times 500/24h, relaxing muscles between lifts—exercises are boring but relieve pain in 80%). Reasonable results are also reported with electrical stimulators. **medline:9208609** If symptoms last >1yr, arthroscopic lateral retinacular release may be tried. If pain still persists, rarely consider patellectomy. Shaving the posterior surface of the patella gives uncertain results.

Excessive lateral pressure syndrome Retropatellar tenderness/pain is felt laterally. Exercise provokes pain. Vastus medialis exercises are less likely to help. The patella is normal. Lateral retinacular release can help.

Bipartite patella This is usually an incidental radiographic finding but may give pain if the superolateral fragment is mobile with tenderness over the junction. Extra fragment excision relieves pain. **medline:8150823**

Recurrent patella subluxation A tight lateral retinaculum causes the patella to sublux laterally giving medial pain. The knee may give way. It is commoner in girls and with valgus knees. It may be familial, or associated with joint laxity, a high-riding patella (*patella alta*), or a hypoplastic lateral femoral condyle. Signs: increased lateral patellar movement, eg accompanied by pain and the reflex contraction of quadriceps (ie a +ve patellar apprehension test). Treatments vary (randomized trials are needed). If vastus medialis exercises fail to help, lateral retinacular release may cure. Patellar tendon transfer is rarely needed. **medline:10981218**

Patella tendinopathy (Jumper's knee) is usually initiated by micro- or macro patella tendon tears, eg associated with sudden sporting loads. It can occur anywhere in the patellar tendon. It settles with rest \pm NSAIDs. For those unable to rest, steroid injection around (not into) the tendon may help. For Osgood–Schlatter's, see p 652. **medline:10323501**

Iliotibial tract syndrome Synovium deep to the iliotibial tract is inflamed as it rubs on the lateral femoral condyle (which is tender). Common in runners, it settles with rest, NSAIDs or steroid injection. **medline:7237678**

Medial shelf syndrome The synovial fold above the medial meniscus is inflamed. Pain is superomedial. There may be brief locking of the knee (mimics a torn meniscus). **Diagnosis:** Arthroscopy. **Treatment:** Rest, NSAIDs, local steroid injection; or division of the synovial fold arthroscopically.

Hoffa's fat pad syndrome Think of this in those with meniscus or ligament-type symptoms when investigation shows they are intact. **MRI** may show a hypertrophic Hoffa pad impinging between the articular surfaces of the joint. This causes pain under the patella. There may be hyarthrosis or haemarthrosis (from arteriole rupture). Extending a bent knee while putting pressure on the patellar tendon margins elicits a strong pain and a defensive behaviour. **medline:9676201**

Results of meta-analyses in knee osteoarthritis

(NB: knee OA is the commonest joint disease in Europe)

- Paracetamol and quadriceps-strengthening exercises are helpful. (EULAR)*
- Weight loss in women ↓ risk of getting OA symptoms. 16 trials
medline:1543306
- Trials comparing NSAIDs are often flawed because the drugs compared tend not to be used in comparable doses, as expressed as a fraction of the maximum daily dose. 16 trials
medline:10796306
- There seems to be little benefit from intra-articular steroid injections beyond a placebo effect (but this is substantial and quite long-lasting). Some trials report benefits which last a week or so. 16 trials
medline:10088770
- Injecting high molecular weight hyaluronan appears to be safe and effective in a meta-analysis done in 1995, but 3 later trials showed no overall benefit at 2–5 months (subgroup analysis shows there may be benefit in those with the severest symptoms). J Chard at
www.clinicalevidence.org
- Transcutaneous electrical nerve stimulation (TENS) for knee osteoarthritis is a noninvasive modality in physiotherapy. Different modes of TENS setting (high rate and strong burst mode) show significant benefit in relief of pain and stiffness over placebo. 7 trials
medline:11034768
- Knee replacement is often the treatment of choice. But 21% of patients are still getting knee pain one or more years post-op. Significant complications include preventable thromboembolism. Intermittent pneumatic compression devices and low molecular weight heparin are significantly better than warfarin and aspirin in preventing DVT. Incidence of asymptomatic PE: 11.7% for aspirin, 8.2% for warfarin, and 6.3% for pneumatic compression group. (Numbers of symptomatic PEs are too small to produce reliable statistics.) medline:10990299
- Post-op aseptic loosening and other problems requiring revision are a problem in 9% over 5yrs for unicompartmental prostheses (7% over 4yrs for bicompartmental prostheses). Serious infections may be less in those centres doing many procedures. J Chard at
www.clinicalevidence.org

*Key reference: EULAR European Task Force 2000 *Ann Rheu Dis* 59 936 [11087696]

Knees that swell

Arthritis Knee swelling may occur with any arthritic process. Osteoarthritis particularly affects the posterior patella and the medial compartment, so tending to varus deformity. Treatment is usually with NSAIDs (p 672), knee support (Tubigrip®), weight loss, local steroid injections, knee replacement (p 654), or osteotomy (corrects varus deformity). If these fail or are unsuitable, some may benefit from making the synovial fluid more lubricant.* Rest pain (or pain while driving) which is relieved by movement suggests patellofemoral compartment problems. Simply pulling the patella medially using tape (Leukotape®, Beiersdorf) may reduce pain by 25%.

Rheumatoid, gout and septic arthritis all affect the knee. Remember that a unilaterally swollen knee may be a septic arthritis. Diagnosis is made on the basis of aspiration, so avoid steroid injections prior to this (p 656).

Meniscal cysts *Typical patient:* A young man. Swelling is variable. Pain may be felt over the joint line. Lateral cysts are commoner than medial. Swelling maximal on knee flexion to ~60° (less noticeable at full flexion). The meniscus is often torn radially (an otherwise unusual direction) so there may be knee clicking and giving way. *Diagnosis.* MRI. *Histology:* Myxoid degeneration. *Treatment* Arthroscopic decompression relieves pain. medline 110322

Ligament tears, meniscus lesions, patellar dislocation See p 710.

Osteochondritis dissecans An osteochondral fragment separates from underlying bone. *Cause:* Unknown. *Typical site:* Lateral side of the medial femoral condyle. *Typical patient:* Young adult/adolescent. *Symptoms.* Pain after exercise with intermittent knee swelling. Locking may occur. *Radiography:* Look for articular surface defects. *Treatment:* None, initially, as spontaneous healing can occur. Attached sections may be excised to prevent it breaking off, or it may be pinned in place—or the fragment can be drilled, in the hope that fibrous tissue formation will secure the fragment, to fill the defect. The condition predisposes to arthritis. medline 108632

Loose bodies These cause knee locking (all movements may be jammed unlike locking from torn menisci when only extension is limited). Also, there is effusion and swelling. *Diagnosis.* Plain films ± ultrasound (helps show if fragments are intra-articular). medline 10442075 *Differential* Tophi (gout). medline 980511 *Causes* Osteochondritis dissecans (up to 3 loose bodies), osteoarthritis (≤10 loose bodies), chip fractures of joint surfaces (≤3 loose bodies) or synovial chondromatosis (>50 loose bodies). When locking is a problem loose bodies are best removed. This can often be done arthroscopically.

Bursitis There are 16 bursae around the knee. The most commonly affected are the prepatellar bursa ('housemaid's knee') where swelling over the anterior inferior patella is due to inflammation and fluid in the bursa due to friction (kneeling); the infrapatellar bursa (Vicar's knee—they kneel more upright), and the semi-membranous bursa in the popliteal fossa (popliteal cyst which differs from the 'Baker's cyst' which is a herniation from the joint synovium). Prepatellar bursae may be aspirated, have hydrocortisone injected to decrease recurrence, or, if very persistent, may need excision. Pain may be relieved by topical NSAIDs. Aspiration distinguishes friction bursitis from infective suppurative bursitis, which needs drainage and antibiotics, eg flucloxacillin 250mg/6h po (adult dose). medline 187204

*This technique involves a 5-week course (2mL/wk) of high molecular weight intra-articular hyaluronan derivatives (eg Synvisc®). Any effusion is drained to dryness. A 5-week course may be repeated at 6 months. Trials are few, but at least some benefit is likely, if all standard alternatives are contra-indicated, e.g. NSAIDs. see *Drug Ther Bull* 1999 37 71, SE. transient pain and knee swelling; synovitis. See p 637

A journey on foot

By the time we get down to the foot, our anatomical interest is flagging, but on close inspection we would never have believed, my dear José, that so much glory could fit into a shoe.¹ Feet made for walking are really quite old (~4–7 million years—according to Sterkfontein Man's remains).² The significant thing about these old feet is that they can and did take us anywhere. We left off swinging from tree to tree in an African forest, and began quite a journey—on foot.

Ankle examination 25° of extension (dorsiflexion) and 30° of flexion is the norm at the tibiotalar joint. Inversion and eversion are effected at the subtalar and midtarsal joints. Toes should have between 60° and 90° extension. Note any callosities. Examine the arches. Watch as the toes are lifted off the ground, and on standing on tiptoe. Examine gait and shoes (normal wear pattern is medially under ball of foot, posterolaterally at heel).

Hallux valgus The big toe deviates laterally at the metatarsophalangeal joint. Biomechanical forces, pointed shoes and wearing of heels forcing the forefoot forward probably promote deformity. Pressure of the metatarsophalangeal joint against the shoe leads to bunion formation. Secondary arthritis in the joint is common. Bunion pads and plastic wedges between great and second toes may relieve pain but severe deformity requires surgery. Many different operations are used. The medial side of the metatarsal head may be trimmed; Keller's operation removes the proximal half of the proximal phalanx (excision arthroplasty) leaving a flail toe; displacement osteotomy of the metatarsal bone; Mayo's operation (arthroplasty excising the distal metatarsal head) and arthrodesis of the MTP joint are all used.

Pes planus (flat feet) The arch of the foot is low. There may be valgus and eversion foot deformity. Flat feet are normal when a child is learning to walk. The medial arch develops over the next few years. If flat feet persists no action is needed if the medial arch restores itself on standing on tiptoe. In most, it is asymptomatic but occasionally it is painful (especially 'peroneal spasmotic flat foot' in which the heel is held everted and the medial border of the foot held flat to the ground. Here, attempts to invert the hind foot induces painful peroneal muscle spasm). Exercises, faradic foot stimulation and medial shoe heel wedges may not yield much benefit. With the spasmotic type, hindfoot fusion can be needed for pain relief. Prevention: studies of feet in India show that going barefoot until 6 years old keeps feet healthy. Pain and limitations in sport tend to be post-op complications (especially after the Viladot procedure), so don't advise surgery lightly. medline:108841

Pes cavus Accentuated longitudinal foot arches may be idiopathic, or from spina bifida or previous polio. Clawed toes may occur, as weight is taken on metatarsal heads when walking. If simple measures fail (padding under the metatarsal heads to relieve pressure), consider surgical soft tissue release (detach ligamentous tissues from calcaneum, manipulate arch then put foot in plaster) or arthrodesis to straighten the toes. If osteoarthritis of the tarsal bones is the cause of pain, these may be fused.

Hammer toes These are extended at the metatarsophalangeal joint, hyperflexed at the PIP joint and extended at the DIP joint. Second toes are most commonly affected. Arthrodesis at the PIP joint straightens the toe. Tenotomy of extensor tendons lets it lie flat.

Claw toes Extended at the metatarsophalangeal joint but flexed at both PIP and DIP joints; this deformity is due to imbalance of flexors and extensors (eg after polio). If toes are still mobile, a Girdlestone operation is needed (a flexor/extensor transfer operation).

¹ Gabriel Gacia Marquez 1989 *The General in His Labyrinth* ² Toumai man is 7×10^6 yrs old.

Problems with hallux valgus surgery

5–33% of patients are dissatisfied at follow-up following Keller's excision and other arthroplasties—even when the hallux valgus angle and pain are improved. So does any one technique confer real benefits? The answer is that there is no compelling evidence, except for evidence against Keller's excision arthroplasty.¹ It may best be reserved for the low-demand patient with symptomatic degenerative changes in the first metatarsophalangeal joint in the absence of lateral metatarsalgia.

Compared with Keller's excision arthroplasty, considerable narrowing of the forefoot is obtained with the modified Mayo operation.² That there is no especial advantage to Keller's arthroplasty is borne out by a Cochrane meta-analysis³—particularly in terms of range of motion and intermetatarsal angle. It is hoped that the newer basal-type osteotomies, such as the proximal crescentic osteotomy, the proximal chevron osteotomy, or the proximal oblique osteotomy combined with distal soft tissue releases will give better long-term outcomes. But we note that 3 trials ($N=205$) have compared chevron-type osteotomy with other techniques—and they had no advantages.

The use of continuous passive motion post-op can give an improved range of motion and earlier recovery. Early weightbearing is not detrimental to final outcome.

1 A Zembsch 2000 *Clin Orthop* **376** 183 [10906874] 2 R Majkowski 1992 *Foot Ankle* **13** 317 [1398359] 3 Cochrane Database Syst Rev 2000 CD000964 [10796404]

The painful foot

► Look at a wet footprint (and wear on the shoe sole) to assess function anatomy of the loaded foot. Many of the conditions mentioned in 'the foot' (p 640) can be painful. Other causes of pain are mentioned here.

Hallux rigidus 1st metatarsophalangeal joint arthritis causes pain and ↓ movement. A dorsal ring of osteophytes may occur. Arthrodesis, Keller's (p 641) or distal oblique osteotomy may be tried. medline:1086231

Young people Children rarely complain of foot pain: if they complain pain in the soles think of a foreign body. Shoe pressure on a prominent navicular bone, or sometimes an accessory bone, or a prominent posterosuperior os calcis may require surgical trimming. Osteochondritis (p 650) of foot bones may be the cause (diagnose by radiography). Köhler's disease involves the navicular bone; Freiberg's disease the metatarsal heads; and Sever's disease is epiphysitis of the heel. Relieve pressure by shoe inserts (or occasionally plaster casts) until pain subsides.

Metatarsus primus varus: 1st metatarsal angulation towards the midline. Typical patients: teenagers. It may be familial. If deformity is great, metatarsal or wedge osteotomy may be tried. medline:1038435


Ingrowing toenails (Typically the big toe.) Incorrect nail cutting: pressure of shoes predispose to the lateral nail digging into its flesh bed, which reacts by becoming heaped up infection-prone 'proud flesh'.




Conservative treatment involves tucking cotton-wool soaked in surgical spirit under the proud flesh and awaiting nail growth (then cut it straight with edges protruding beyond flesh margins). Antibiotics may help the young. medline:9483428 Recurrent infections may require surgery. **Simple nail avulsion** plus **phenol** to the nail bed is better than **simple excision** without use of phenol (but there may be more post-op infection). meta-analysis medline:1079680

Adult forefoot pain (metatarsalgia) Increased pressure on the metatarsal heads causes pain. **Treat** by insole supports. Treatment by surgery other than for rheumatoid arthritis is unpredictable. Other causes of metatarsalgia include synovitis, sesamoid fracture, and injury.

Morton's metatarsalgia Pain is from pressure on an interdigital neuroma between the metatarsals (eg from fashion shoes). Pain usually radiates to the lateral side of one toe, and the medial side of its neighbour (eg toes 3 & 4). A compression test of the affected web space is quite specific, diagnostically. Neuroma excision may be needed. medline:1075101

March (stress) fractures occur in the shaft of the 2nd or 3rd metatarsals and may follow excessive walking. Radiographs may be normal (radionuclide bone scans are more discriminating). Treatment is expectant. If pain is severe, a plaster cast while awaiting healing may help.

Pain in the heel **Causes:**  = may respond to steroid injections)

- Diseases of the calcaneum
- Arthritis of the subtalar joint
- Rupture of calcaneal tendon (p 660)
- Calcaneal paratendinopathy 
- Postcalcaneal bursitis (back of heel)
- Tender heel pad 
- Plantar fasciitis* (pain under anterior calcaneum) 

MRI may have a rôle in undiagnosed heel pain (eg from occult stress fractures). medline 10423942 Apart from calcaneal diseases and tendon rupture conservative treatment (eg shoe alteration to prevent rubbing) may help. If not, with postcalcaneal bursitis the bursa may be surgically removed.

proximal plantar fasciitis doesn't respond to injections, extracorporeal shockwave therapy is a reasonable nonoperative alternative to surgery—2002 meta-analysis [@11991474]

Osteomyelitis

This is an infection of the bone. Its incidence reduces as living standard rise. Common organisms: *Staph aureus*, streptococci, *E coli*, *Proteus* and *Pseudomonas*. Elsewhere: *Salmonella* (with sickle-cell disease) and *Brucella*. Infection may spread from boils, abscesses, pneumonia, or from genitourinary instrumentation. Often no primary site is found. The patient may have diabetes or otherwise impaired immunity. *Other risk factors*: surgical prostheses, open fractures, penetrating injury. medline:1077199

The patient Pain—with tenderness, warmth, and erythema at the affected part; unwillingness to move; slight effusion in neighbouring joints; signs of systemic infection. All signs are less marked in adults.

Cancellous bone is typically affected in adults; in children, vascular bone is most affected (eg in long bone metaphyses—esp. distal femur, upper tibia). This leads to cortex erosion, with holes (cloacae). The pus lifts up the periosteum causing death of the original bone (sequestrum). Continued bone formation by the elevated periosteum forms an involucrum. Pus may discharge into joint spaces or via sinuses to skin.

Brodie's abscess: A less virulent abscess, surrounded by sclerotic bone (the 'halo' on MRI); drain, and give antibiotics for ~6 weeks. medline:8324315

Tests ESR/CRP↑, WCC↑. Blood culture (+ve in 60%). MRI is sensitive and specific (88% and 93%, vs 61% and 33% for isotope scans)—and avoids ionizing radiation. But isotope scans are still sometimes needed.* Radiographic changes are not apparent for a few days but then show haziness ± loss of density of affected bone, then subperiosteal reaction, and later, sequestrum and involucrum. Infected cancellous bone shows less change.

Treatment Drain abscesses and remove sequestra by open surgery. Culture all sequestra. Antibiotic: flucloxacillin 0.5–1g/6h ivi until the organism and its sensitivities are known. Continue for 6 weeks. Alternative treatments for adults are fusidic acid or clindamycin. Ciprofloxacin 500mg/8–12h po is suitable for *Pseudomonas* osteomyelitis. In children <5yrs, *Haemophilus influenzae* may be the cause, and ampicillin may be appropriate—but be guided by sensitivities, and a microbiologist.

Complications of osteomyelitis Septic arthritis; fractures; deformity.

Chronic osteomyelitis Poor treatment results in pain, fever, and sinus suppuration with ± long remissions. Radiographs show thick irregular bone. Excise sequestra and 'saucerize' cavities. Hyperbaric oxygen may be tried. UK chambers tel 02392 822351 Give antibiotics (as above) for ≥12 weeks.

Complications: Amyloid, squamous carcinoma development in sinus track.

Bone TB (eg vertebral body) 1–3% of all TB; incidence is rising, rare in UK.

Spread: Haematogenous or via nearby nodes. **Signs** Local pain, swelling, and 'cold abscess' formation ± joint effusion are common. With joint involvement there is pain, swelling, pain on movement and muscle wasting. Also: weight↓; malaise; fever; lethargy. **Differential**: Malignancy; other infections; gout; rheumatoid. **Imaging**: Radiographic changes: bone rarefaction, periostitis changes, cyst formation. Later: loss of joint space, erosions, bony ankylosis. Also look for soft tissue swellings (abscess; tenosynovitis/bursitis). MRI is especially useful in analysing soft tissue changes. Bone scans may be useful in diagnosing dactylitis (a feature of childhood TB). **Other tests**: Aspirated pus is creamy and may be positive on culture or ZN stain. ESR↑, Mantoux +ve. CXR. HIV tests. **Treatment**: Drain abscesses, immobilize affected large joints. Standard 6-month courses (OHCM p 568) of eg isoniazid (300mg/day), rifampicin (600mg/day) and pyrazinamide (1.5g/day) may not be long enough. Late arthrodesis is needed for gross joint destruction.

medline:9182026

Bone tumours

Tumours present with pain, swelling or pathological fracture. Bone is common site for secondaries (eg from breast, bronchus, prostate, kidney). Primary bone neoplasia is rare: 6 million population/year. They all present with non-mechanical pain (like toothache) followed by swelling. Delays in diagnosis are common. Radiological features suggestive of bone tumour include: bone destruction, new bone formation, soft tissue swelling, and periosteal elevation. Metastases are blood-borne and usually arise first in the lungs. Treatment of these rare and highly aggressive tumours is best carried out in multidisciplinary specialist treatment centres.

Giant cell tumour (osteoclastoma) This histologically 'benign', primary skeletal neoplasm with an unpredictable pattern of biological aggressiveness is commonest in young adults. It occurs around epiphyses (especially the knee). The tumour is osteolytic and often slowly progressive, sometimes resulting in pathological fracture. Treatment is by detailed and thorough curettage resulting in 75% cure. Recurrences are usually treated by wide excision. Metastasis is rare.

Ewing's sarcoma This malignant round-cell tumour of long bones (typically diaphysis) and limb girdles, usually presents in adolescents. Radiographs show bone destruction, concentric layers of new bone formation ('onion-peel' sign) and a soft tissue mass. MRI is helpful. Typically those with Ewing's sarcoma have a t(11;22) chromosomal translocation. Chemotherapy, bone resection, and limb salvage surgery or radiotherapy cures ~half. The key adverse prognostic factor is metastases at diagnosis (5-year recurrence-free survival is 22%—vs 55% if no metastases at diagnosis).

medline:1096363

Osteosarcoma Primary osteosarcoma typically affects adolescents and arises towards the end of long bones (typically the knee), in the metaphysis. Secondary osteosarcomas may arise in bone affected by Paget's disease or after irradiation. **Radiography:** Bone destruction and new bone formation, often with marked periosteal elevation (sunray spiculation and Codman's triangle respectively). **Treatment:** Resection + chemotherapy, eg doxorubicin and cisplatin in 3-week cycles for 18 weeks (safer, and as good as multi-drug regimens). **ECM** 9314864 A cure rate of ~50% is achievable. Vigorous treatment of metastatic disease may be worthwhile.

Chondrosarcoma may arise *de novo* or from malignant transformation of chondromas. It often presents on the anterior chest wall arising from the costochondral arches or sternum. It is usually associated with pain and presents in the axial skeleton of the middle-aged. 'Cotton wool calcification' is typical on radiography. MRI/CT will better define tumour extent. They don't respond to chemotherapy or radiotherapy, so treatment is by excision. Inadequate surgery is accompanied by local recurrence, often of a higher grade of malignancy. The cure rate is ~70%.

medline 1045125

Malignant fibrous histiocytoma (The rarest primary bone tumour) arise in middle age, often at the site of previous bone abnormality (eg bone infarcts). Treatment is as for osteosarcoma. mcb.uni-bonn.de/cancernet/207864.html

Chordomas arise from notochord remnants in the spine and are typically sacral, and quite big before a diagnosis is made. Bladder & bowel sphincter problems may occur. Radical surgery + pre-op radiotherapy cure <20%. The others may have a slow and difficult death. medline:11104990

Surgical reconstruction (after excising a bone tumour) involves replacing affected bone with a metal and polyethylene endoprosthesis—

an alternative to amputation. Excellent and durable reconstruction is possible using massive endoprostheses or bone allografts. 85% of patients now have limb salvage following chemotherapy for primary bone tumours.

Benign tumours *Osteoid osteoma* occur most commonly in long bones of males 10–25yrs old, and cause severe pain. They appear as local cortical sclerosis on radiographs. **Treatment:** Local excision. **NB:** In young adults, any bone pain responding to aspirin within 15min could be caused by an osteoid osteoma. A radionuclear scan can confirm or exclude.

Chondroma: These benign cartilaginous tumours may arise from bone surfaces or within the medulla (enchondromata). They may cause local swelling or fracture. **Treatment** is rarely needed, unless, for example, the acromioclavicular joint is involved. **NB:** Carney's syndrome is gastric leiomyosarcoma, plus pulmonary chondroma plus a non-adrenal paraganglioma. medline:865533

Osteochondroma: (Autosomal dominant) This commonest type of benign tumour. See *Hereditary multiple exostoses*, p 648.

Fibrous dysplasia of bone: See McCune–Albright syndrome, p 754.

Soft tissue sarcomas unrelated to bone (**NB:** osteosarcomas and Ewing's tumours may be extraskeletal.) Whenever an unexplained lump presents, eg in a limb, think: could this be a sarcoma (ie a mesenchymal tumour of connective-tissue elements)? ▶ *Muscle sarcomas comprise 1% of all malignancies.* **MRI** is the image of choice. http://rpisun1.mda.uth.tmc.edu/se/sts/

Possible diagnoses: rhabdomyosarcoma; leiomyosarcoma, synovial sarcomas, neurofibrosarcoma; fibrosarcoma; liposarcoma; malignant or pyogenic histiocytoma; haemangiopericytoma. Gene expression profiling has been used for classification and prediction of response to chemotherapy (usually only to doxorubicin and ifosfamide). * If *not* a sarcoma, is it a metastasis, lymphoma, distended bursa, pseudotumour, haematoma, or an infective or granulomatous diseases (Wegener's; sarcoid; TB; actinomycosis; toxoplasmosis; amoebiasis; cysticercosis; pyodermitid), or cystic lesion? Simple shelling-out of the sarcoma is **not** wise: refer to a tertiary centre skilled in pre-op assessment—particularly if the lesion is >5cm across or deep-seated. medline:9657781 & 8154283

Chemotherapy and metastectomy have a rôle. 5-year survival of children with rhabdomyosarcoma is now approaching 80%. medline:10754991

Congenital disorders of bone & cartilage*

Osteogenesis imperfecta (Adair Dighton syndrome) A genetic disorder of collagen causing brittle, fragile bones. There are 4 types:

- I Blue sclerae with deafness but little deformity (defective formation of type I collagen; autosomal dominant).
- II Lethal perinatal form with many fractures and dwarfism.
- III Fractures at birth + progressive deformity; blue or white scleral hue; dentinogenesis imperfecta common (enamel separates from defective dentine, leaving teeth transparent or discoloured; recessive).
- IV Fragile bones, white sclerae after infancy; autosomal dominant.

Radiographs: Many fractures, osteoporotic bones with thin cortex, and bowing deformity of long bones. **Histology:** Immature unorganized bone with abnormal cortex. **Treatment:** Prevent injury. Osteotomies may correct deformity. Intramedullary rods are sometimes used in long bones.

Achondroplasia Reduced growth of cartilaginous bone (membranous bone growth is OK) from mutations eg in the fibroblast growth factor receptor 3 (FGFR3) gene. Affected babies have large heads, normal trunk length, short limbs and fingers all the same length. Adults are dwarfed with ↑lumbar lordosis, bow legs, and shortened proximal arms and legs. **Radiographs:** Short bones with wide epiphyses. Growth hormone has been tried. medline.10509364

Craniocleidodysostosis Those affected have a flat face, a globular skull, absent clavicles ± wide perineum (if coxa vara). **Radiographs:** Midline frontal bone suture; deficient clavicles; wide pubic symphysis; coxa vara. Coxa vara is correctable, if needed, by osteotomy. Life expectancy is normal.

Hereditary multiple exostoses (HME diaphyseal aclasis) is a common-ish autosomal dominant disorder in which certain proteins accumulate in the Golgi apparatus leading to cartilage-capped tumours (exostoses/osteochondromata) developing from affected cartilage nests at the end of long bones. These point away from the near-by joint. If severe, bones are broad and badly modelled, causing short stature as well as forearm, knee, and ankle deformity. Beware of malignant transformation to chondrosarcomas** or osteosarcomas. HME is usually caused by defects in various EXT genes which encode enzymes catalyzing biosynthesis of heparan sulfate, an important component of the extracellular matrix. Although genetic linkage analysis has identified 3 loci at 8q24, 11p1113, and 19p (genes EXT1, EXT2, & EXT3), most HME is attributed to missense or frameshift mutations in genes encoding 746- and 718-aa proteins, which are expressed in many human tissues. **Treatment:** Removal of symptom-producing exostoses. medline 10639137

Ollier's (Maffucci's) enchondromatosis is a nonhereditary disease characterized by multiple central (medullary) cartilaginous radiolucent bone tumours of unknown pathogenesis. It usually involves the extremities with a unilateral predominance. Sarcoma transformation may occur. Small bones of hands, feet, upper tibia, and lower femur are chiefly affected. Whole-body MRI detects associated haemangiomas & gliomas. medline 10664698

Osteopetrosis Lack of differentiation between cortex and medulla of bone results in very hard dense 'marble' bones that are brittle. Anaemia and thrombocytopenia may result from decreased marrow space. Deafness and optic atrophy can result from compression of cranial nerves. Lack of remodelling preserves variations of ossific density causing the characteristic 'bone within a bone' appearance. www.sxiagram.com, margaret/338 1552 1641440 jpg

Developmental bone biology

Because bone is ossified, we tend to think of it as the architectural rock around which our living tissues are constructed. But bone maintenance and development is a highly dynamic and regulated process sensitive to a wide variety of hormones, inflammatory mediators, growth factors, and genetic influences which become aberrant whenever there are deletions, insertions, and missense mutations. medline:10529789 The concept of a master gene is useful here—to indicate how genes relate and interact. Master genes encode proteins that can control other genes by directly binding to their DNA. For example, transcription factor *Osf2* (osteoblast specific transcription factor 2) gene is thought to serve as a master gene regulating expression of other genes, allowing mesenchymal stem cells to differentiate into osteoblasts. *Osf2* maps close to a chromosomal locus on chromosome 6p21 (thought to be important eg in cleidocranial dysostosis). NB: master genes make a mockery of genes vs environment issues. One gene is an environment for another, and the effects of each may be catastrophic in some environments or negligible in others. www.bbc.co.uk/science/horizon/hopefulmonsterstran.shtml

See also Neurofibromatosis (OHCM p 390), Marfan's (OHCM p 730), Ehlers–Danlos & Morquio's (p 746).
Cartilage tumour classification: Is the lesion benign or malignant? Is the lesion a pure or impure cartilaginous tumour? Is the epicentre of the lesion intraosseous, juxtacortical, or in the soft tissues? The most common benign tumours are enchondroma, osteochondroma, chondroblastoma, and chondromyxoid fibroma. Chondrosarcoma is malignant. [@8446748]

Osteochondritis

Osteochondritis juvenilis (osteochondrosis) is a condition of unknown origin in which bony centres of children or adolescents become temporarily softened, resulting in deformation secondary to pressure. After a variable period of time (often about 2 or 3 years) the bone hardens again in its new, deformed shape.

The Patient The age at which these changes occur depends on which bone is affected, as it is usually at the time of development of the bony nucleus. The main symptom is local pain. *Radiographs:* Initial increase in density of the bony nucleus; followed by patchy appearances (as affected bone is resorbed) and flattening; then normal texture returns but deformity is maintained. The long-term consequence is degenerative arthritis.

Name of osteochondritis

Perthe's disease (p 630)

Scheuermann's disease

Köhler's disease

Kienböck's disease

Freiberg's disease

Site affected

Hip

Vertebral ring epiphyses

Navicular bone

Lunate bone (in adults)

Head of 2nd or 3rd metatarsal

Treatment Often none is necessary but large joints may need protection (eg with a plaster cast).

Scheuermann's kyphosis This is the commonest cause of kyphosis in 13–16-yr-olds. It is an autosomal dominant. The normal ossification of ring epiphyses of several thoracic vertebrae is affected. Deforming forces are greatest at their anterior border, so vertebrae are narrower here, causing kyphosis. During the active phase, vertebrae may be tender. *Radiographs:* Irregular vertebral endplates, Schmorl's nodes and ↓ disc space ± anterior wedging. Schmorl's nodes are herniations of the intervertebral disc through the vertebral end-plate. *Treatment:* If posture control (eg standing during lessons rather than sitting) and exercises (eg swimming) fails, physiotherapy ± spinal braces can help. Surgery may be tried for severe kyphosis (>75°) with curve progression, refractory pain or neurologic deficit. medline 10084088 & <http://web.idirect.com/~xray/ced/ced00079.htm>

Köhler's disease Children affected are 3–5-yr-olds. Pain is felt in the mid-tarsal region and they limp. There may be navicular tenderness. *Radiographs:* Dense, deformed bone. *Treatment:* Symptomatic: resting the foot or wearing a walking plaster. *Prognosis:* Excellent. medline 7993505

Kienböck's disease Pain is felt over the lunate (esp. during active wrist movement). Grip is impaired due to pain. *Radiographs:* Dense lunate with a little depth reduction early; more marked flattening later. Osteoarthritis is a consequence. *Treatment:* Early: try wrist splinting for 3 months; later: lunate excision with prosthesis implantation. Once arthritis is established lunate excision does not help. www.cid.ch/TEACH/AF/AF08.htm

Freiberg's infarction This may be classed as an osteochondritis dissecans (p 652), eg of the 2nd metatarsal head, presenting as forefoot pain around puberty, in girls. There may be microfractures at the junction of the metaphysis and the growth plate. *Radiographs:* Epiphysis of a metatarsal head becomes granular, fragmented and flattened. *Treatment:* Good shoes ± metatarsal pad. Limit activity for 4–6 weeks. If severe consider removal of affected bone with bone grafting or interpositional arthroplasty and use of a walking plaster. www.medmedia.com/o11/201.htm

Osteochondritis-like conditions

Calvé's vertebra (Like Scheuermann's disease, p 650, but rarer.) Typical site: a thoracic vertebra (if neck or atlas, torticollis may result). Histology: Look for eosinophilic granulomata. Typical age: 2–10yrs. Symptoms: Pain and \pm tenderness over the affected vertebra; slight kyphosis.

Radiographs/CT/MRI: Dense, flattened vertebral body with disc space preserved. **Bone scan:** Hyperactive spot. $\Delta\Delta$: Sarcoma; osteomyelitis.

Treatment: Bed rest \pm plaster-cast immobilization. www.txt.co.uk/full.cfm?1081

Osgood–Schlatter's disease This tibial tuberosity apophysitis and quadriceps tendonitis affects children 10–14yrs old. $\sigma/\text{f} \approx 3:1$. The Pain below the knee is worse on strenuous activity and quadriceps contraction (lift straight leg against resistance). The tuberosity looks big, and is tender.

Radiographs: Tibial tuberosity enlargement (\pm fragmentation). **NB:** diagnosis is clinical, not simply radiological. The cause may be strain on the developing tuberosity. MRI shows the tendonitis.

Treatment: Rest. Some advocate ice compresses. www.txt.co.uk/full.cfm?1080

Plaster cast immobilization for 6 weeks may be needed. Tibial tubercle excision has been recommended if the above fail. medline:7790481

Sinding Larsen's disease Traction tendinopathy with calcification in the proximal attachment of the patellar tendon, which may be partially avulsed. Symptoms, treatment are similar to Osgood–Schlatter's disease (above), but the onset tends to be 1–2yrs earlier. medline:721864

Sever's disease This common calcaneal apophysitis is probably from strained attachment of the Achilles tendon. It is usually self-limiting. Typical age: 8–13yrs. There is pain behind the heel (bilateral in 60%) \pm limping, and tenderness over the lower posterior calcaneal tuberosity.

Radiographs: Often normal.

Treatment: Physiotherapy. If needed, a below knee walking plaster may give pain relief. Most are well after 5 weeks. medline:3793908

Osteochondritis dissecans See p 638 for knee osteochondritis. Other sites: elbow, hip, ankle, talus. A loose body is formed (1–3cm across) from a convex joint surface when a segment of subchondral bone and cartilage becomes avascular and separates from underlying bone. Adolescents or young adults experience early aching and effusions after use, and sudden painful locking of joints once pieces have separated to make loose bodies.

Radiographs. Look for lucent areas in a piece about to separate, the defect from which the piece has separated, and loose bodies after separation.

Treatment: Expectant \pm removal of loose bodies. Closed wedge resection may lead to revascularization (eg capitellum). medline:10921645

Avascular necrosis (osteonecrosis) **Sites of infarction:** Hip (commonest), knee, shoulder. **Imaging** (MRI is best) Sclerotic or porotic bone due to infarction; joint surface odd, osteochondral fragments. medline:10875132

Local causes: Trauma (eg fractured neck of femur); secondary to rheumatoid, severe osteoarthritis, psoriatic arthropathy, or neuropathic joints.

Systemic: Thalassaemia, sickle-cell (+ any cause of microthrombi, eg platelets \uparrow in leukaemia), NSAIDs/steroids (eg post-transplant), SLE, scleroderma, SBE, dyslipidaemia, alcoholism, pancreatitis, diabetes, big burns, radiation, diving accidents (p 802), Cushing's & Gaucher's diseases. medline:11013361

Treatment Immobilization; analgesia; for hips, arthroplasty. medline:10925752

Joint replacement

Joint replacement has been used for 30yrs. Each year 40,000 hips and 15–20,000 knees are replaced in the UK (knees are overtaking hips).

Hip replacement The usual indication for operation is pain (surgery for fixed flexion deformity affecting walking is less successful). 75% are to replace osteoarthritic hips. These patients are usually elderly. Rheumatoid arthritis (RA) is the next most common indication (patients are often in their 30s or 40s). Other conditions which may result in replacement are avascular necrosis of head of femur; congenitally dislocated hip; fracture neck of femur.

Many prostheses are available but most consist of a metal femoral component with an intramedullary stem usually held in place by bone cement and a plastic acetabular component. Early success of operation occurs in 90%. Early complications include: dislocation (0.5–3%) and deep infection (<1%). Deep infection is a disaster and although exchange prosthesis using antibiotic-loaded cement may be possible, a Girdlestone excisional arthroplasty may be needed. A small proportion of implants fail to make a satisfactory interface with bone.

Later problems of loosening or infection are heralded by return of pain. Plain radiographs are inconclusive in the case of loosening, strontium or technetium scans may reveal increased bone activity. Suspected sepsis should be investigated by white cell count, ESR, and gallium scan. Scans are not reliable within 8 months of operation. Revision arthroplasty is more successful for loosening than for infection. By 9–10yrs post-op 11% of implants have been revised. Be cautious in recommending replacement to those in their 60s who are likely to cause excessive wear of the prosthesis. Excess weight should be lost as this also contributes to usage. Earlier replacement is used for rheumatoid arthritis as joints tend to be grossly affected younger—and excessive delay may result in surgery upon very rarefied osteoporotic bone.

Central migration of the prosthesis via perforation of the medial acetabular wall is a rare cause of external iliac artery & bladder injury. meta-analysis
medline:11869708

Knee replacement The knee is more complicated than the hip, and designs for replacements have altered greatly, from long-stemmed hinge mechanisms, to short-stemmed articulations currently used. Indications for knee replacement: pain at rest, or disturbing sleep, or making house-bound. Pain correlates poorly with radiological signs. Success rate: 95%. Joint survival: 90% last 15yrs (better than hips). Revision rates are similar. Quality of life can be transformed, even if >80yrs old. www.txt.co.uk/full/cfm?1069

Other joints Joint spacers are used in finger joints for rheumatoid, with success. Elbow replacements are beginning to show some success. Shoulder replacement success rates are approaching those of knees.

Preventing thromboses DVT occurs in $>2/3$ of major orthopaedic events, but fatal PE in only 0.1–0.2%. medline:8950998 Low-molecular weight heparin (LMWH) halves DVT rate and lowers risk of fatal pulmonary embolus (PE) by $2/3$. In major orthopaedic surgery, low-molecular weight heparin (LMWH) is preferable to ordinary heparin. ci: uncontrolled bleeding/risk of bleeding (eg peptic ulcer); endocarditis; children. *Dose example:* dalteparin 2500u 2h pre-op and at 12h post-op, then 5000u once/day for 1 week. In knees, fondaparinux 2.5mg/day may be better than enoxaparin. medline:11794149 DVT/PE prophylaxis may need continuing for 1 month post-op. NB: if the rate of fatal PE is low (~0.1%), in some centres prophylaxis may not be warranted. medline:7822392

Infected hip and knee prostheses

This may be a disastrous complication of joint replacement—presenting with pain, fever, or ESR↑. Sinus formation may relieve pockets of pus. Gallium scintigraphy may help make the diagnosis. Get expert help from radiologists, an orthopaedic surgeon specializing in the problem, and a consultant in infectious diseases.

Treatment: Early on, debridement + antibiotics may be enough. medline:877205 Later, with loosening of components, radical debridement must include removal of all prosthetic material, as well as any involved bone and soft tissue. Sometimes reconstruction by exchange arthroplasty works, eg for less virulent infections. Antibiotics may be needed for months. Patient selection is vital, eg those without draining sinuses, without immunocompromise, and with adequate bone quality after debridement. medline:10611868 The Mayo Clinic reports a 13% failure rate with a 2-stage reconstruction. Another alternative may be the removal of all foreign material and allowing ankylosis to occur, ie a Girdlestone pseudarthrosis, for the hip.

SBE-style antibiotic prophylaxis ± antibiotic-impregnated cement? **Dentistry-associated prosthesis infection** with oral bacteria is very rare and risks of antibiotics are palpable (>500 penicillin-caused deaths/yr, in the USA), and SBE-style prophylaxis is not recommended. BNF 2002 **Colonoscopy + polypectomy** may be more risky than dentistry; some recommend prophylaxis, if <6 months since replacement. medline:8772051 The best prophylaxis might be achieved with a combination of gentamicin-impregnated cement (on its own, unreliable), systemic antibiotics for >24h, and surgical enclosure. However, the additional cost of the surgical enclosure may prove too expensive in some areas. Reimplantation need not require use of cement: hydroxyapatite-coated total prostheses are an alternative. medline:8772051 & 10366918

Joint injection and aspiration

Joint aspirations *Diagnostic rôle:* Any blood, crystals, or pus? 2002 meta-analysis medline:1200632
Therapeutic rôle: For tense effusions, septic effusions, and haemarthroses

Steroid injections to inflamed joints, bursae, or tendon sheaths aim to ↓inflammation and relieve pain, perhaps by ↓prostaglandin synthesis, stabilizing mast cells, or ↓tissue calcification, or increasing vascularization and permeability of synovium. *Side-effects:* Skin atrophy (hydrocortisone acetate is safer than triamcinolone), haemarthrosis, facial flushing, urticaria, Charcot's arthropathy, post-injection flare syndrome (synovitis with fever), paresis, and septic arthritis (≤ 1 in 14,000 injections). www.clinicalevidence.org
▶It is essential that steroids are not used in septic conditions and, if any doubt at all exists, results of synovial fluid culture should be awaited. Remember the possibility of tuberculous synovitis—especially in immigrant populations. Repeated injections are more dangerous: beware ligamentous laxity, joint instability, calcification, or rupture of tendons.

Preparations are available: *hydrocortisone acetate* (cheapest, shortest acting), *methylprednisolone*, and *triamcinolone*. They may be mixed with 1% lidocaine. When triamcinolone is used for injecting near short tendons 10mg strength is preferred to 40mg as tendon rupture has been reported after the latter. Despite our best intentions 'joint' injections often fail to meet their target (50% in one study in which contrast material was also injected); those off-target are less likely to relieve symptoms. medline:825788

Conditions responding reasonably well to steroid injection
Localized subdeltoid bursitis; supraspinatus, infraspinatus, and subscapular tendinopathy; shoulder arthritis; tennis and golfer's elbow, arthritis of elbow, radioulnar, acromioclavicular, and sternoclavicular joints; ganglia; trigger fingers; strains of collateral and cruciate ligaments of knee; suprapatellar, infrapatellar, and Achilles tendinopathy; plantar fasciitis; traumatic arthritis of metatarsophalangeal joints; and sesamo-first-metatarsal joint.

Preparation Check that you have swabs, needles, and sterile bottles. For aspiration of viscid fluid (eg haemarthrosis) use 19G needle. For the large joints use a 21G (green, in the NHS) needle, and for fingers and toes a 23G (blue) needle. Locate joint margins carefully before cleansing with chlorhexidine in 5% spirit or surgical spirit, and once the skin is cleansed use scrupulous aseptic no-touch technique. Remember that antiseptic and local anaesthetics take 3min to work; even then, the skin is clean but not sterile. Samples for microbiology should be sent in sterile containers; those for cytology or crystal examination in heparinized or FBC containers.

Knee joint (▶see p 637) The patient lies with knee supported slightly flexed and muscles relaxed. Palpate the joint space behind patella either medially or laterally. Insert a needle horizontally between the patella and femur. Slight resistance is felt on traversing the synovial membrane; it should be possible to aspirate fluid, and injection fluid should flow easily.
Usual doses: 50mg hydrocortisone acetate, 40mg methylprednisolone, 20mg triamcinolone hexacetonide. Repeat injections should be longer than 3 months apart. If injection is used for prepatellar bursitis, give 25mg hydrocortisone acetate into the most tender spot.

The ankle Plantar flex foot slightly, palpate joint margin between extensor hallucis longus (lateral) and tibialis anterior (medial) tendons just above tip of medial malleolus. Inject 25mg hydrocortisone acetate into the joint.

Synovial fluid in health and disease

Aspiration of synovial fluid is used primarily to look for infectious or crystal (gout and pseudogout) arthropathies.

	Appearance	Viscosity	WBC/mL	Neutrophils
Normal	Clear, colourless	High	<200	<25%
Noninflammatory ¹	Clear, straw	High	<5000	<25%
Haemorrhagic ²	Bloody, xanthochromic	Variable	<10,000	<50%
Acute inflammatory ³	Turbid, yellow	Decreased		
Acute gout			~14,000	~80%
Rheumatic fever			~18,000	~50%
Rheumatoid arthritis			~16,000	~65%
Septic	Turbid, yellow	Decreased		
TB			~24,000	~70%
Gonorrhoeal			~14,000	~60%
Septic (non-gonococcal) ⁴			~65,000	~95%

eg, degenerative joint disease, trauma.

eg, tumour, haemophilia, trauma.

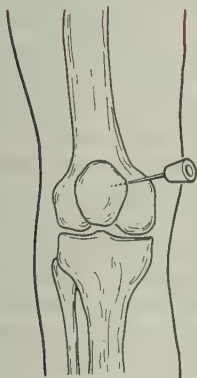
includes eg Reiter's, pseudogout, SLE etc.

includes staphs, streps, Lyme, and *Pseudomonas* (eg post-op).

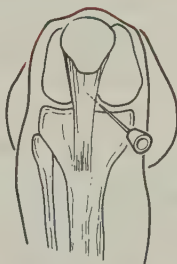
For inflammatory causes of arthritis: Synovial fluid WBC >2000/mL is 84% sensitive (84% specific); synovial fluid neutrophil count >75% is 75% sensitive (92% specific). ►NB: not all labs are equally skillful.

meta-analysis
medline:12006320

(a)



(b)



The right knee, extended (anteromedial approach).

The right knee, flexed (infrapatellar anteromedial approach).

Produced by kind permission from M Crawley. *Br J Hosp Med* 11 747-55

Joint injections—continued

Shoulder injection Because shoulder pain from soft tissue causes a common (lifetime incidence: ~10%), and because the pain can be long-lasting (only 23% resolve within a month), this is one of the most commonly injected joints, but we do not know *who* to inject or *when* in their illness, because trials are few, and the best ones cast doubt on the benefit of *any* injection.

Anterior approach: (for aspiration, synovitis, frozen shoulder) seat the patient with arm relaxed by side of his chest. Feel the space between head of humerus and glenoid cap about 1cm below coracoid process. Insert 21-gauge green needle into the joint space (enters joint space when almost up to its hilt) and inject 25–50mg hydrocortisone acetate. Be sure not to go medial to the coracoid process. **Lateral approach:** (subacromial bursitis, painful arc syndrome) inject 25–50mg hydrocortisone acetate with lidocaine just below lateral tip of acromion, pointed downwards and advanced medially. When the needle is withdrawn from touching the head of humerus with slight pressure on the plunger, a drop in pressure is felt as the bursa is entered. Painful arc pain may be reproduced. A second injection may be given after >48h.

Tennis elbow 25mg hydrocortisone acetate with 1mL lidocaine is injected with force to area of maximal tenderness over lateral humeral condyle, moving to and fro down to bone several times. A second injection may be needed, eg after 2 weeks. Warn the patient that symptoms may worsen briefly after the injection has been given. (Avoid triamcinolone and Depo-Medrone®: as injections are superficial, fat necrosis may occur.)

Elbow joint injection With elbow flexed at 90°, inject 25mg hydrocortisone acetate between proximal head of radius (locate by rotating patient's hand) and lateral epicondyle by lateral approach (needle 90° to skin), or posteriorly between olecranon and lateral epicondyle.

Biceps tendinopathy is worsened by externally rotating the arm. Insert needle parallel to tendon (if resistance, it is in the tendon: withdraw a bit) and inject 25mg hydrocortisone acetate into tendon sheath and 25mg into joint.

Wrist injection Inject 25mg hydrocortisone acetate 1–1.5cm deep between extensor tendons of ring and little fingers between ulnar head and lunate.

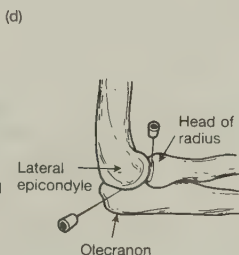
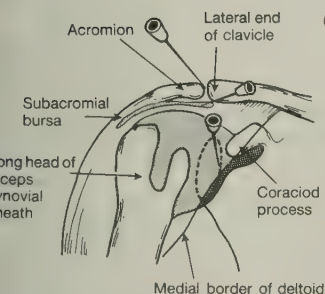
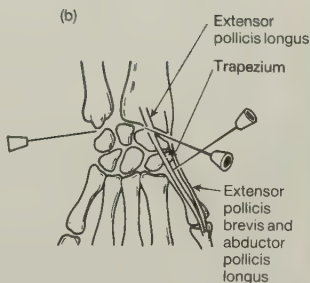
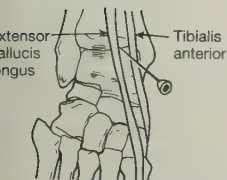
De Quervain's tenosynovitis Extensor pollicis brevis and abductor pollicis longus tendons, eg on traversing the extensor retinaculum on the dorsal wrist, may cause a tender swelling (p 616). With needle almost parallel to skin pointing proximally, inject 25mg hydrocortisone acetate slowly just distal or proximal to the radial styloid, at the site of maximum tenderness. If needle in tendon, injection is difficult so withdraw until easy flow occurs.

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Carpal tunnel Place the hand comfortably palm up. Inject 25mg hydrocortisone acetate at 90° through the distal transverse skin crease of the palmar surface of the wrist closer to the hamate than the trapezium—ie a few mm to the ulnar side flexor carpi radialis. Other techniques are also used. Sometimes the needle enters the median nerve causing sudden pain in the fingers—so reposition the needle. If the first injection fails, consider repeating with 10–20mg triamcinolone. A splint worn for the next few days may mitigate symptoms which can occur at the time of injection.

Trigger finger Insert needle at proximal finger skin crease parallel to flexor tendon, pointing towards palm. Palpate tendon thickening in palm and proceed as for De Quervain's.

First carpometacarpal joint of thumb Avoiding radial artery, inject 25mg hydrocortisone acetate at base of first metacarpal at 1cm depth in anatomical snuffbox (aim at base of little finger).



Dorsal aspect of the right ankle indicating anatomical landmarks.
 Dorsal aspect of the right wrist indicating anatomical landmarks.
 Diagrammatic representation of the anterior aspect of the right shoulder region. The stippled areas indicate synovial membranes of the subacromial bursa and the glenohumeral joints.
 The right elbow, flexed.

Produced by kind permission from M Crawley.

Br J Hosp Med 11 747

Tendon rupture and injury

The main tendons to rupture are the extensor tendons of the fingers, the Achilles tendon, the long head of biceps (p 612), supraspinatus (p 612), and the quadriceps expansion. The cause may be sharp or blunt trauma (anything from sporting injuries to rubber bullets). Lancet 1999; 353: 179

Ultrasound aids diagnosis (its usefulness is largely operator-dependent).

Ruptured extensor tendons of the fingers The long extensors inserting into the distal phalanges are particularly prone to rupture by trauma resulting in 'mallet' or 'baseball' fingers. The affected digit is splinted for 3–6 weeks (in extension). If the tendon doesn't reattach to bone spontaneously, the choice is between living with the deformity or operative repair. The long extensor of the thumb may rupture as a complication of fractures to the lower radius. **Treatment:** Repair uses the lower end of the extensor indicis tendon re-routed to be sutured to the distal thumb tendon (as the thumb extensor is too frayed for direct repair). The extensors of the ring and little fingers are particularly prone to spontaneous rupture in rheumatoid arthritis (attrition rupture from the distal ulna).

Ruptured Achilles (calcaneal) tendon Sudden pain is felt at the back of the ankle during running or jumping as the tendon ruptures ~5cm above the tendon insertion. Pain may be perceived as a kick. It is still possible to walk (with a limp), and some plantar flexion of the foot remains but it is impossible to raise the heel from the floor when standing on the affected leg. A gap may be palpated in the tendon course (particularly within 24h of injury). On examination he cannot stand on tiptoe.

The squeeze test (Simmonds' test) is sensitive: ask the patient to kneel on a chair, while you squeeze both calves—if the Achilles is ruptured there will be less plantar flexion on the affected side. **Treatment:** Urgent tendon repair (most can be percutaneous rather than open) is often preferred to conservative treatment (immobilization of the lower leg in plaster with the foot slightly in equinus). Undiagnosed ruptures may repair, but tendon lengthening occurs. medline:1098671

Ruptured quadriceps expansion Injury may be direct (eg a blow) or indirect (stumbling causing sudden contraction of the apparatus). The quadriceps expansion encloses the patella and inserts into the tibial tuberosity as the patellar tendon. **Non-traumatic causes:** Pseudogout, Wilson's disease; renal failure with hyperparathyroidism. medline:1088753

Treatment: Rupture can occur at the site of quadriceps insertion to the patella, through the patella by fracture (if reduction is possible it is secured by wire or screws, if not, the patella is excised and tendon repair effected), or by avulsion of the patellar tendon from the tibial tuberosity (in young patients the tear may be resutured). After repairs a plaster cylinder is worn for several weeks, then intensive physiotherapy is needed to regain knee function. Many have persistent weakness and difficulty returning to higher level sporting activities. medline:961946

Tenosynovitis Tendons and surrounding synovium may become locally inflamed (possibly from strain) thereby causing pain, eg supraspinatus tendinopathy and bicipital tendinopathy (p 612), or De Quervain's (p 616). Acute frictional synovitis at the wrist is another example, with swelling over the wrist and thumb extensors. If palpation doesn't detect crepitus, at some sites, stethoscope auscultation reveals sounds like bronchial breathing.*

Treatment: In wrist tenosynovitis, a splint for 3 weeks (leaving the finger free) may be needed to allow inflammation to subside.

*R Adler 1999 *NEJM* 340 156 [9917225]—'Bronchial breathing' on the painful right arch on great toe flexion with 'vesicular'-type breathing on the unaffected left side

Nerve compression syndromes

For brachial plexus and peripheral nerve lesions, see p 724, 664 & 720.

Carpal tunnel syndrome ▶ This is the commonest cause of hand pain at night. It is due to compression of the median nerve as it passes under the flexor retinaculum. Pregnancy, the Pill, myxoedema, lunate fracture (rare), rheumatoid arthritis, cardiac failure, or being premenstrual may increase compression. *Clinical features* are of median nerve distribution:

- Tingling or pain are felt in the thumb, index finger, and middle finger.
- When the pain is at its worse, the patient characteristically flicks or shakes the wrist to bring about relief. Pain is especially common at night and after repetitive actions. Affected persons may experience clumsiness. Early on there may be no signs. Later there is:
- Wasting of the thenar eminence & ↓ sensation over the lateral 3½ fingers
- Lateral palmar sensation is spared as its supply (the palmar cutaneous branch of the median nerve) does not pass through the tunnel.
- Phalen's test: holding the wrist hyperflexed for 1–2min reproduces the symptoms. (This is more reliable than Tinel's test—tapping over the tunnel to produce paraesthesiae. Note: Phalen's flexing, Tinel's tapping.)

Tests: Nerve conduction studies are not usually needed (and may be –ve)

Treatment: Carpal injection: p 658. (Symptom relief beyond one month compared to placebo is unproven.) medline:11034724 Wrist splints worn at night may relieve nocturnal pain. More permanent results are obtained from decompression by flexor retinaculum division.

Ulnar nerve compression at the elbow This is described on p 614.

Radial tunnel syndrome (Posterior interosseus nerve compression)

This radial nerve branch is sometimes compressed on passing through the supinator muscle (eg after forearm fracture or excessive exercise). Patients experience weakness of the thumb and fingers. *Examination* may reveal weakness of long finger extensors, and short and long extensors of the thumb, but no sensory loss. *Treatment* after trauma is surgical decompression and sprung splints to extend fingers. medline:785200

Anterior interosseus nerve compression This median nerve branch may be compressed under the fibrous origin of flexor digitorum sublimis causing weakness of pinch and pain along the forearm's radial border. Examination shows weakness of the long thumb flexor and flexor profundus to the index and middle fingers. *Treatment* is surgical decompression.

Ulnar nerve compression at the wrist Uncommon. Loss may be motor or sensory. *Diagnose* by electrophysiology. *Treat* by surgical decompression.

Meralgia paraesthetica If the lateral cutaneous nerve of the thigh is compressed (on leaving the pelvis just medial to the anterior superior iliac spine, eg by tight jeans), pain and paraesthesiae may be felt over the upper outer thigh. Sensation may be ↓ over this area. It is usually self-limiting, and can occur in pregnancy. *Treatment:* Cortisone and local anaesthetic injection at the anterior superior iliac spine gives unpredictable results. Complete or partial pain relief was found in 72% after decompression and in 82% after transection of the nerve. medline:756627

Common peroneal compression Nerve compression against the head of fibula (eg plaster casts, thin patients lying unconscious, proximal fibula fracture, squatting, obstructed labour) causes inability to dorsiflex the foot. Sensation may be ↓ over the dorsum of the foot. *Test* Electrophysiology. *Treatment:* Most recover spontaneously but surgical decompression may be needed (eg if 3 months elapse without improvement). Physiotherapy and splint until foot-drop recovers. medline:1074105

Soft tissue injuries

Correct management of soft tissue injuries reduces pain, recovery time and subsequent disability.

Treatment in the first 24 hours: 'RICE'

Rest: A splint or plaster cast may help.

Ice: Cold is anaesthetic and a vasoconstrictor. Apply ice packs (eg a packet of frozen peas wrapped in a cloth) or cold compresses intermittently for <10min at a time, to avoid cold injury.

Compression: Strapping restricts swelling and further bleeding.

Elevation: Ideally elevate the affected part above the heart to improve drainage from the affected part and to reduce pain.

Rehabilitation Passive stretching to maintain joint mobility and muscle length, then progressive active exercise until the full range and strength of movement is restored, eg wobbleboards for ankles. Sportsmen must then train to full fitness. Also consider NSAIDs, rubefacients, and ultrasound.

Sports injuries Many sports injuries can be prevented.

Is the patient preparing the body for activity with a proper warm-up? Inadequate warm-up increases the risk of injury.

Cooling-down is also important in reducing muscle soreness.

Is protective equipment being worn?

Many acute and chronic injuries are caused by unsuitable equipment, faulty technique, or unwise training schedules—advice from a professional coach can be invaluable.

Common patterns of injury in sport: 'Shin splints'—shin soreness, common in unfit runners on hard surfaces and due to muscle tears, mild anterior compartment syndrome or stress fracture; knee pain—most causes on p 636; ligamentous ankle injury (p 712); plantar fasciitis (p 642); stress fractures such as march fracture—suspect with increasing bony pain despite normal radiographs (radionuclide scans are more discriminating); Achilles tendon problems; tennis elbow (p 614).^[a]

Overuse phenomena

Overuse phenomena at work (Work-related upper limb injury = occupational overuse syndrome = isometric contraction myopathy = repetitive strain injury =

RSI) Activity requiring repetitive actions, particularly those associated with prolonged muscle contraction, may lead to chronic symptoms. Employers have a duty to provide a safe working environment and well-designed chairs and tools, and frequent short breaks. Changes of posture and activity help to reduce work-related upper limb injury (this term is preferred to repetitive strain injury). The cost of these injuries in suffering, and hours lost from work, is considerable as treatment of established symptoms is often difficult and may necessitate change in employment—if one is available.

Compensation is a vexed issue, and recent Court judgments have gone in favour of the employers in some instances, and in favour of patients in others. Some people argue that the condition does not exist as a separate medical entity emphasizing lack of histopathology. It should be noted that this is not a prerequisite for a disease (see *Sudden infant death*, p 286)—and in any case histopathology is sometimes demonstrable.^[a] Treatments tried include rest (may prolong the problem), physiotherapy, β -blockers for relaxation, and the Alexander technique for posture re-education.

Those who use vacuum cleaners, assemble cars, or play stringed instruments may all develop overuse phenomena.

Testing peripheral nerve motor functions

Nerve root Muscle

C3,4	trapezius
C4,5	rhomboids
C5,6,7	serratus anterior
C5,6	pectoralis major (clavicular head)
C6,7,8	pectoralis major (sternocostal head)
C5,6	supraspinatus
C5,6	infraspinatus
C6,7,8	latissimus dorsi
C5,6	biceps
C5,6	deltoid

Test—by asking the patient to:

Shrug shoulder (via accessory nerve)
 Brace shoulder back.
 Push arm forward against resistance.
 Adduct arm from above horizontal, and push it forward.
 Adduct arm below horizontal.
 Abduct arm the first 15°.
 Externally rotate arm, elbow at side.
 Adduct arm from horizontal position.
 Flex supinated forearm.
 Abduct arm between 15° and 90°.

The radial nerve

C6,7,8	triceps	Extend elbow against resistance.
C5,6	brachioradialis	Flex elbow with forearm half way between pronation and supination.
C5,6	extensor carpi radialis longus	Extend wrist to radial side with fingers extended.
C6,7	supinator	Arm by side, resist hand pronation.
C7,8	extensor digitorum	Keep fingers extended at MCP joint.
C7,8	extensor carpi ulnaris	Extend wrist to ulnar side.
C7,8	abductor pollicis longus	Abduct thumb at 90° to palm.
C7,8	extensor pollicis brevis	Extend thumb at MCP joint.
C7,8	extensor pollicis longus	Resist thumb flexion at IP joint.

Median nerve

C6,7	pronator teres	Keep arm pronated against resistance.
C6,7	flexor carpi radialis	Flex wrist towards radial side.
C7,8,T1	flexor digitorum sublimis	Resist extension at PIP joint (while you fix his proximal phalanx).
C8,T1	flexor digitorum profundus I & II	Resist extension at the DIP joint of index finger.
C8,T1	flexor pollicis longus	Resist thumb extension at interphalangeal joint (fix proximal phalanx).
C8,T1	abductor pollicis brevis	Abduct thumb (nail at 90° to palm).
C8,T1	opponens pollicis	Thumb touches base of 5 th finger-tip nail parallel to palm).
C8,T1	1 st and 2 nd lumbricals	Extend PIP joint against resistance with MCP joint held hyperextended.

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Ulnar nerve

C7,8,T1	flexor carpi ulnaris	Flex wrist towards ulnar side.
C7,C8	flexor digitorum profundus III and IV	Fix middle phalanx of little finger, resist extension of distal phalanx.
C8,T1	dorsal interossei	Abduct fingers (use index finger).
C8,T1	palmar interossei	Adduct fingers (use index finger).
C8,T1	adductor pollicis	Adduct thumb (nail at 90° to palm).
C8,T1	abductor digiti minimi	Abduct little finger.
C8,T1	flexor digiti minimi	Flex the little finger at MCP joint.

The musculocutaneous nerve (C5–6) This may be injured at the brachial plexus, causing weakness of biceps, coracobrachialis, and brachialis. Forearm flexion is weak. There may be some loss of sensation.

	Muscle	Test—by asking the patient to:
S1, S2	gluteus medius and minimum (superior gluteal nerve)	Internal rotation at hip, hip abduction.
S1, S2	gluteus maximus (inferior gluteal nerve)	Extension at hip (lie prone).
S3, S4	adductors (obturator nerve)	Adduct leg against resistance.

peroneal nerve

S2, S3	iliopsoas	Flex hip with knee flexed and lower leg supported: patient lies on back.
S3	sartorius	Flex knee with hip external rotated.
S3, S4	quadriceps femoris	Extend knee against resistance.

obturator nerve

S3, S4	Hip adductors	Adduct the leg.
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superior gluteal nerve

S1, S2	Gluteus maximus	www.rad.washington.edu/atlas2/gluteusmaximus.html Hip extension.
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superior gluteal nerve

S5, S1	Gluteus medius	www.rad.washington.edu/atlas2/gluteusmedius.html Abduction and internal rotation of hip.
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peroneal nerve (*including the common peroneal nerve)

S1, S2	hamstrings	Flex knee against resistance.
S5	tibialis posterior	Invert plantarflexed foot.
S5	tibialis anterior	Dorsiflex ankle.
S1	extensor digitorum long	Dorsiflex toes against resistance.
S1	extensor hallucis long.	Dorsiflex hallux against resistance.
S1	peroneus longus & brevis	Evert foot against resistance.
S1	extensor digit. brevis	Dorsiflex hallux (muscle of foot).
S1	gastrocnemius	Plantarflex ankle joint.
S1, S2	flexor digitorum longus	Flex terminal joints of toes.
S1	small muscles of foot	Make sole of foot into a cup.

Quick screening test for muscle power

Shoulder	Abduction C5	Hip	Flexion	L1-2
	Adduction C5-7		Extension	L5-S1
Elbow	Flexion C5-6	Knee	Flexion	S1
	Extension C7		Extension	L3-4
Wrist	Flexion C7-8	Ankle	Dorsiflexion	L4
	Extension C7		Plantarflexion	S1-2
Fingers	Flexion C7-8; Extension C7			
	Abduction T1			

The UK MRC scale objectifies strength (reasonably well):

Grade 0	No muscle contraction	Grade 3	Active movement against gravity
Grade 1	Flicker of contraction	Grade 4	Active movement against resistance
Grade 2	Some active movement	Grade 5	Normal power

Grade 4-, 4, and 4+ describes movement against slight, moderate, and strong resistance. Dynamometers help quantify strength. medline:11773212

Remember to test proximal muscle power: ask the patient to sit from lying, pull you towards himself, and to rise from squatting. ▶ Also, observe walking (easy to forget, even if the complaint is of walking difficulty!)

Root numbers in **bold** indicate that that root is more important than its neighbour. Differences vary in ascribing particular nerve roots to muscles—and there is some biological variation in individuals. The above is a reasonable compromise, and is based on the MRC guidelines.

Assessing the locomotor system

We should measure things not by appearance but by function.*Senneca ~65A
With her arthritic fingers, can she do up zips or buttons? Can she open a tin? Is she using any special devices to assist in daily life? What can't she do that she would like? Does she have any comments on her joints? (Details of testing each muscle may be found on p 664.)

Essence Ask questions, look, compare, move, and palpate. If a joint feels normal to the patient, looks normal, and has a full movement, it usually is normal.

Valgus or varus? In a valgus deformity, remember 'L' for lateral in the spelling: the part of the limb distal to the deformity is angled laterally from the midline. Varus is the other way.

3 screening questions •Are you free of pain or stiffness in your muscles? •Can you dress all right? •Can you manage stairs? If 'Yes' to all 3, muscle problems are unlikely. If 'No' to any, go into functional detail.

A screening examination To be done in light underwear. (No corsets!)

Observe from behind: Is muscle bulk ok (buttocks, shoulders)? Is the spine straight? Symmetrical paraspinal muscles? Any swellings or deformities?

Observe from the side: Normal cervical and lumbar lordosis? Kyphosis?

'Touch your toes, please': Are lumbar spine and hip flexion normal?

Observe from in front for the rest of the examination. Ask him to:

'Tilt head towards shoulders': Is lateral cervical flexion normal?

Open and close your mouth': Orofacial pain, temporomandibular joint noise: ± restricted jaw function indicate temporomandibular joint problems, p 534

'Put hands behind head': Tests glenohumeral and sternoclavicular movement

'Arms straight': Tests elbow extension. Then test supination and pronation

Examine the hands: See OHCM p 37. Any deformity, wasting, or swellings?

'Put index finger on thumb': Tests pincer grip. Assess dexterity.

Observe the legs: Normal quadriceps bulk? Any swelling or deformity?

Find any knee effusion: Sit the patient down, take the leg on your lap, and do the patella tap test (this also tests for patellofemoral tenderness), or to be more sensitive, watch any fluid moving from compartment to compartment by stroking upwards over the medial side of the knee, and then downwards over the lateral side. Aspirate it. Any blood, crystals, or pus?

Observe feet: Any deformity? Are arches high or low? Any callosities? These may reflect an abnormal gait of some chronicity.

'Walk over there please': Is the gait smooth? Is there good arm swing? Is the stride length ok? Normal heel strike and toe off? Can he turn quickly?

Additional palpations •Midpoint of trapezius and the medial end of supraspinatus muscle, to elicit the tenderness of fibromyalgia (p 675).

•Squeeze across 2nd–5th metacarpals. Any synovitis? Repeat in metatarsals.

•Palpate for crepitus with your palm on his knee (patient supine), while you passively flex knee and hip to the full extent. Is movement limited?

•Internally rotate each hip in flexion.

The GALS system for quickly recording your findings*

G (Gait) ✓

Approved by the Arthritis & Rheumatism Council

Appearance: Movement:

A (Arms)

✓

✓

L (Legs)

✓

✓

S (Spine)

✓

✓

A tick (✓) means normal. If not normal, then substitute a cross with a footnote to explain what the exact problem is.

*M Forstater & V Radin 2001 *The Spiritual Teachings of Seneca*, Hodder, p 109

Hot, swollen joints

The term arthritis implies an inflammatory condition of one or more joints. Features of inflammation: *pain, swelling, overlying redness, stiffness, and local heat* (\pm fever). In making a diagnosis, there are 3 key questions:

- Is only one joint affected (ie a monoarthritis)?
- Is the patient otherwise well, or is there a disease causing arthritis (eg ulcerative colitis) or a risk factor for infection (diabetes, renal failure, immunosuppression, iv drug abusers)?
- What is making the joint swollen? Blood, crystals or pus?—It is always worth aspirating the joint (p 656) to find out, but beware introducing infection if the effusion is a sympathetic one, from nearby cellulitis or abscess formation.

Note: the presence of rheumatoid factor (latex +ve, p 670) divides the arthritides into seropositive (sp below) and seronegative.

Causes Any polyarthritis can start as a monoarthritis. This list is only a guide to likely presentation. ► Always think of infection—eg TB.

Monoarthritis:

- Bacteria, eg staphs, streps, pseudomonas, gonococci; TB; septicaemia
- Gout or pseudogout
- Reiter's disease (OHCM p 406)
- Traumatic haemarthrosis
- Psoriasis
- Rheumatoid arthritis^{SP}
- Leukaemic joint deposits
- Reactive arthritis (see p 675)

Polyarthritis:

- Viruses, eg rubella, mumps, Hepatitis A, Epstein-Barr
- Rheumatoid arthritis^{SP}
- Sjögren's syndrome^{SP}
- Rheumatic fever
- Systemic lupus erythematosus^{SP}
- Ulcerative colitis; Crohn's
- Drug reaction
- Reiter's and psoriasis (rarer)

^{SP}=seropositive for rheumatoid factor

Tests Liaise with a microbiologist; take blood cultures before treatment.

- Gram stain, culture, and microscopy (for crystals, using polarized light) on fluid aspirated from the joint. Blood staining of synovial fluid is seen in pseudogout as well as after trauma, but it is rare in septic arthritis.
- Radiography of the part. Look for signs of trauma (fractures), rheumatoid arthritis (p 670). Cartilage calcification suggests pseudogout. In some patients radiographs will appear normal.
- Urate, FBC, ESR, Latex, C-reactive protein (normal <10mg/L) may be \uparrow in sero +ve or -ve causes; it does *not* screen for sepsis. Consider HIV serology.

Management Unless microbiological facilities are readily to hand, consider hospitalizing all people with acute monoarthritis of a large joint so that infection can either be ruled out, or appropriate antibiotics given. Even if aspiration reveals pus, it does not follow that infection is the cause (the diagnosis may be pseudogout or rheumatoid arthritis).

Treatment is determined by cause. In septic arthritis, give flucloxacillin (for staphs—adult: $\frac{1}{2}$ –1g/6h slowly iv; child: 25mg/kg/6h) + benzylpenicillin 1.2g/4h iv until sensitivities are known. In infants, *Haemophilus* is common so give cefotaxime too (50mg/kg/12h iv slowly). Look for atypical mycobacteria and fungi if HIV +ve. Ask a microbiologist how long to continue treatment (eg 2 weeks iv then 4 weeks of oral therapy). NB: the rôle of repeated aspiration, lavage, and surgery is uncertain. Ask an orthopaedic surgeon's advice. Many will recommend repeated arthroscopy for a child's infected hip. Beware inviting in new pathogens.

Splints help by immobilizing the joint in the early days of treatment. If a joint prosthesis is present, surgery with removal of all foreign material may be required. Vigorous physiotherapy follows antibiotics.

Osteoarthritis (OA)

For knee OA, see p 636–7

OA is a disease of synovial joints with degeneration and destruction of hyaline cartilage. Predisposing causes: trauma, joint malalignment (eg valgus deformity); ↓size of joint contact (eg subluxation in congenital dislocations); osteochondral fractures; foreign bodies; torn menisci; damaged cartilage from septic arthritis or neuropathy. Early on there is splitting (fibrillation) of cartilage, then some breaks away, leaving exposed bone—which becomes thickened, hard, and white ('eburnation'). Nodular new bone formations at joint surfaces are called osteophytes.

The patient may suffer joint pain at rest (eg at night); pain on movement. Joints may be a little thickened, but not usually inflamed. Crepitus (the grating of surfaces) may be felt on joint movement. There may be fixed deformity (inability to assume neutral position). In the hands the distal interphalangeal joints are commonly affected. Swellings at these joints are called Heberden's nodes. Rheumatoid factor is absent. The ESR is normal.

Radiographic changes 1 Joint space ↓ 2 Subchondral sclerosis 3 Subchondral cysts 4 'Lipping' at joint margins (from osteophytes).

Treatment Aim to reduce pain and disability, and to increase fitness and quality of life. Gentle-to-moderate exercise improves symptoms, but note that cycling may aggravate patellofemoral osteoarthritis, and swimming is generally good for back and hip symptoms but may worsen cervical apophyseal joint osteoarthritis. There are no panaceas. Local warmth may give relief. Physiotherapy may increase functional use of joints. Unload weight-bearing joints through weight reduction (diet) and walking aids (stick, frame, stair rail). Occupational therapists can advise on modifying tools and home. Intra-articular steroid injections are also tried, and may relieve symptoms for a month or two (eg in the knee: see p 656). For pain, try paracetamol 500mg–1g/6h po alone or in combination with opioids, eg co-proxamol (expect constipation). Consider intermittent NSAIDs, eg ibuprofen 400mg/8h po pc, for inflammatory episodes (eg knee), but if needed long term, beware GI bleeding, and try a misoprostol-containing tablet, eg Arthrotec® (contains diclofenac), or Napratec® (contains naproxen). Good data about which NSAID is best are scarce. medline:10796384

When disability or pain are severe, consider surgery. Knee OA is the commonest joint disease in Europe: see p 637 for detailed interventions.

Surgery Arthroscopic joint debridement (± excision of osteophytes and unstable articular cartilage ± drilling of exposed bone) helps ≥60%.

Arthrodesis (surgical fusing of a joint) is used most commonly at the metatarsophalangeal joint of the hallux.

Osteotomy to realign a joint—eg to correct genu varum deformity. Excision arthroplasty is used in Keller's operation (p 641, for problems).

Interposition arthroplasty is used at the thumb's carpometacarpal joint, when a silastic block is interposed between the joint surfaces.

Joint replacement (p 654) for hips, knees, elbows, and shoulders.

Arthritis of the hip Fixed flexion deformities are common. Thomas test reveals such deformity (p 628). The usual surgical treatment is replacement arthroplasty (p 654).

Neuropathic joints (Charcot's joints)

Causes Diabetic neuropathy, tabes dorsalis, cauda equina lesion, leprosy, syringomyelia (upper limb). Gross disorganization of joints ± dislocation follows minor traumas or overstretching unconstrained by pain.



The abnormal left hip has a narrowed joint space, a result of loss of the articular cartilage. An osteophyte protrudes laterally from the acetabular roof, the sub-articular bone is sclerotic, there is cyst formation, and the head of the femur is distorted. These are the changes of osteoarthritis. The lateral shift of the femoral head suggests that the underlying cause is congenital subluxation of the hip.

NSAIDs and cyclo-oxygenase (COX-1 & COX-2) medline:9929039

NSAIDs suppress cyclo-oxygenase enzyme (cox) which converts arachidonic acid to prostaglandins—so producing analgesic and anti-inflammatory effects. cox exists in 2 forms: cox-1 which mediates gastric protection and maintains platelet function. cox-2 is undetectable in most normal tissue but its expression increases in inflammation (ie it is inducible) and it produces the prostaglandins which mediate inflammation. cox-2 inhibitors such as rofecoxib produce analgesic and anti-inflammatory effects comparable to non-selective NSAIDs, with claimed (be sceptical!) less risk of peptic ulcer. They are not true 'gastroprotectors', and they are still problematic in those with peptic ulcers, and they may cause problems with fluid retention, as cox-2 is expressed in the kidney's medulla densa. They don't have aspirin's cardiovascular effects: on the contrary, thrombosis is increased (relative risk \uparrow 2–3-fold) by their propensity to \downarrow vasodilatory and antiaggregatory prostacyclin production. medline:11509060

Nevertheless cox-2 inhibitors add another option, eg if there is a history of peptic ulcer, or there is high risk of developing a peptic ulcer (eg elderly, debilitated, or requiring maximum doses of NSAID over prolonged periods—or taking other agents likely to promote bleeding eg: rofecoxib 12.5–25mg/24h po). www.nice.org.uk

Rheumatoid arthritis (RA)

The central immunological event is said to be presentation of the culprit antigen (whatever this turns out to be) to T-helper cells, with subsequent cytokine-mediated synovial neutrophil exudate, which releases cartilage-degrading enzymes. There is a symmetrical synovitis, with arthralgia, stiffness, and swelling, and, if prolonged, destruction of peripheral joints.

Prevalence 3% of females, 1% of males. Common in middle age (but any age affected). Association: HLA-DRW4. Incidence: 1/1000 adults/yr.

Presentation Very variable. It may take time before RA can be distinguished from other inflammatory conditions. Onset is often insidious with malaise, transient pain, stiffness, and weakness, but may be sudden—'One day I couldn't move'. Monoarthritis, palindromic arthritis—joints affected by turn—and systemic presentations occur.

Prognosis 20% have one attack, often acute, with complete recovery. 30% are mildly disabled; 40% have prolonged, moderately disabling disease, severe in 10%. Life expectancy↓, by 3 (♀) to 7yrs (♂). Worse prognosis if: female, palindromic onset, extra-articular features, nodules, erosions occurring within 3yrs of onset, persistently high CRP, rheumatoid factor ↑↑, HLA DR4 +ve.

Joint involvement Inflamed synovium (pannus) produces hot swollen tender joints and thickened tendons with associated soft-tissue swelling, hence the characteristic sausage-shaped fingers. Early morning stiffness may last for several hours. With time the pannus erodes the joints and tendons rupture and the irreversible changes typical of advanced RA are seen.

Fingers develop 'swan neck' (hyperextended PIP joint but flexed DIP joint) or 'boutonnière' deformities (flexed PIP joint, extended MCP joint, hyperextended DIP joint); **thumbs** adopt Z-deformities. **MCP joints** and **wrists** sublux acquiring ulnar deviation: the ulnar styloid and distal radius become prominent. **Knees** develop valgus or varus deformity. Popliteal 'Baker's' cysts occur and may rupture mimicking a DVT. In the **feet** subluxation of the metatarsal heads with associated hallux valgus, clawed toes, and callouses causes pain—'walking on marbles'. ▶**Atlantoaxial instability** may lead to cord compression; radiography prior to intubation is wise.

Extra-articular involvement Weight↓, fever, and malaise are common. Many systemic manifestations of RA are caused by vasculitis or nodules. 10% develop amyloid. Drug complications can affect most systems (p 672).

Skin: Nail fold infarcts, ulcers, rashes, palmar erythema.

Nodules: Firm, mobile, non-tender nodules are found on pressure areas, eg forearm of 20% of sufferers. Nodules may also occur on tendon sheaths (causing triggering) or in lungs, myocardium, pericardium, or sclera.

Eyes: Keratoconjunctivitis sicca (p 482), scleritis, episcleritis.

Cardiovascular system: Pericardial effusion, pericarditis, myocarditis.

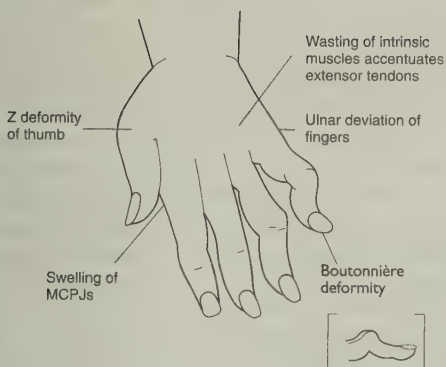
Lungs: Pleurisy, pleural effusions, nodules, fibrosing alveolitis.

Nerves: Entrapment neuropathy (eg carpal tunnel).

Tests **Blood:** Normochromic anaemia, ESR↑. Both correlate with disease activity. Also: alk phos↑, platelets↑, lymphocytes↓, wcc↓ (in Felty's *OHCM* p 724). Rheumatoid factor: this anti-IgG autoantibody appears eventually in 80% of those with RA but is also found in low titres in many chronic conditions. Antinuclear factor: +ve in 30% with RA (*OHCM* p 409).

Synovial fluid Turbid; viscosity↓; clots, from high polymorph count.

Radiographic changes. In order of appearance: soft-tissue swelling; juxta-articular osteoporosis; symmetrical loss of joint space; erosions at joint margins, subluxation, and dislocation of joints.



Wrist subluxation, prominent distal radius.



Rheumatoid arthritis (RA): treatment

► Refer all patients to a rheumatologist as early as possible as vigorous prompt use of disease-modifying drugs can prevent future problems.¹ The patients' attitude is crucial to wellbeing. A severely afflicted patient who does quads exercises daily and seeks ways around disability will be living an independent life long after a mildly affected patient who has 'given up'. **Symptom-control** may require bed-rest (acute flare-ups), NSAIDs* ± passive then active exercise to maintain mobility and muscle tone. HRT may particularly help women patients. **Chronic disease** is characterized by pain, stiffness, malaise and permanent loss of function. Exercises, splints, and gadgets (eg kettle-tippers) protect joints. Special shoes (or trainers) and seat raisers allow independence. **Skeletal collapse** needs analgesia, surgery, appliances/home conversion, and social/financial help to mitigate isolation and income loss.

Disease-modifying antirheumatic drugs (DMARDs)² modify immunology—and ↓ disease activity (ESR ↓, 'tender joint count' ↓, ↑ function, eg grip). As they are slow to act and have serious adverse effects, regular monitoring is vital. Sulfasalazine and methotrexate are commonly-used DMARDs.¹ It is probably the case that all patients diagnosed with RA should be offered DMARDs.

Sulfasalazine 500mg/6h po (start with 500mg/24h) is less effective than IM gold but works faster and is well tolerated, so may be used earlier. SE: D&V, sperm count ↓, wcc ↓. Monitor LFT + FBC. Warn to stop if sore throat (=wcc ↓).

Methotrexate Do LFT, FBC, U&E, hepatitis B & C serology (biopsy liver if +ve, or if alcohol abuse). * Warn to report any cough or dyspnoea (SE: pneumonitis). Stop if wcc $<4 \times 10^9/L$; neutrophils $<2 \times 10^9/L$; platelets $<150 \times 10^9/L$. Methotrexate (eg 7.5mg/week po) with high-dose prednisolone (eg 60mg/24h po tapered in weekly steps to 7.5mg/day over 6 weeks) may have a rôle here. In one study (N=155) this regimen combined with sulfasalazine (2g/day po) was able to suppress disease in 72% compared with 49% in the placebo group receiving sulfasalazine alone. The benefits only lasted while the treatment was being given (prednisolone was tapered and stopped by week 28, and methotrexate by week 40); 6 months after stopping the treatment, CRP, a useful marker of disease activity, was similar in both groups.

Gold (Sodium aurothiomalate 50mg/week IM or, a bit less effective, orally as auranofin 3mg/12h). After 3 months on IM gold 80% of patients improve and can use a lower dose but 60% stop due to side-effects. SE necessitating stopping treatment: mouth ulcers, rashes (risk of exfoliative dermatitis), persistent proteinuria, lung fibrosis, wcc $<4 \times 10^9/L$; neutrophils $<2 \times 10^9/L$; platelets $<150 \times 10^9/L$. ► Monitor urine and FBC often.

Penicillamine up to 500mg/24h po. Efficacy (and monitoring) as for gold.

Hydroxychloroquine 200mg/24h po is least effective. SE: retinopathy so get an ophthalmic opinion before starting, or if any eye symptoms or ↓ acuity.

Immunosuppressants (azathioprine, leflunomide) may be used. medline:1184043

Steroids can reduce erosions if given early (prednisolone 7.5mg/day po). It is not known if all should receive this. One problem is ↓ bone density over long periods. Another problem is that it would be difficult to keep patients to $<7.5mg/day$, as, for the first months, there may be great symptomatic improvement, which then tails off, leaving patients wanting higher doses and risking cataracts, fluid retention, and peptic ulcers.

Surgery may relieve pain and improve function, eg arthroplasty (joint reconstruction/replacement), particularly for forefoot, hips, and knees; arthrodesis (joint fusion) of ankle or foot. Synovectomy, tendon repair and transfer, joint debridement, and nerve decompression are also useful.

Biological response modifiers (BRM)^{2,3,4}

The chief biological event is cytokine production, particularly interleukin-1 (IL-1) and tumour necrosis factor (TNF- α). IL-1 mediates synovial inflammation (pannus), synoviocyte proliferation, cartilage degradation, and bone erosion.³ Long-term results of escalation therapy (first an NSAID, then disease-modifying drugs in increasing doses) give disappointing results, and erosions are prevented if the cells producing the inflammatory mediators are inactivated early.

Anticytokine therapy *Infliximab* (anti-TNF antibody) and *etanercept* (TNF receptors, eg 25mg sc twice-weekly) \downarrow TNF activity, and, in ~40% of those with persistently active disease, activity may be halved (vs 3% on methotrexate alone).^{5,6} SE: few; long-term safety issues are opaque (? \uparrow risk of cancer \pm infections \pm demyelination**). Anticytokines are approved by NICE^{org.uk} in active RA (judged by certain activity measures) who have failed to respond to \geq 2 DMARDs one of which is methotrexate.

Anakinra (eg 100mg/24h sc) is a new IL-1 receptor antagonist mirroring the action of endogenous IL-1 receptor antagonist that is often deficient in RA. It is licensed as an add-on, if methotrexate alone is insufficient.^{7,8}

Does this person have active RA? With these new therapies, this is clearly a critical question. One objective measure is the **28 joint disease activity score**.*** According to NICE, if DAS28 >5.1 (on two occasions separated by 1 month), anticytokines should be considered.

Examples: *ibuprofen* 400mg/8h po pc. *Diclofenac* 25–50mg/8h po pc; it can be combined with misoprostol (Arthrotec®)—an ulcer-protecting agent. Which NSAID will help most is unpredictable. A good response shows inflammation is present—so a DMARD may help. Cox-2 inhibitors have a rôle (p 669). In one 12-week study, etoricoxib 90mg/24h po was about as good as naproxen 500mg/12h po. CI asthma, urticaria, current peptic ulcer, LFT \uparrow , creatinine \uparrow . SE GI bleeding, BP \uparrow , LFT \uparrow . medline-12033987 & www.nice.org.uk

*Pending long-term data, avoid anticytokines if pre-existing multiple sclerosis—discontinue at once if CNS signs or symptoms occur; get expert advice. medline-11762947

**DAS28 = $0.56 \times \sqrt{\text{TJC}} + 0.28 \times \sqrt{\text{SJC}} + 0.7 \times \ln \text{ESR} + 0.014 \times \text{GHS}$

TC is the tender joint count; SJC is the swollen joint count and GHS is the general health status 100mm visual analogue score. Automatic calculators are available to ease the number crunching. See www.das-score.nl/www.das-score.nl/downloads.htm

High disease activity >5.1 , low disease activity <3.2 , remission <2.6 .

M Green 1999 *Rep Rhe Dis* 18 2 P Emery 1997 *Lancet* 350 304 3 A Gough 1994 *Lancet* 344 23 M Boers 1997 *Lancet* 350 309 5 J O'Dell 1999 *NEJM* 340 310 6 M Weinblatt 1999 *NEJM* 340 253 J Jenkins 2002 *Am J Med Sc* 323 197 [at12003375] 8 S Cohen 2002 *Arth Rhe* 46 614 [at11920396]

The seronegative and crystal arthropathies

Diseases causing seronegative arthritides Psoriasis, Reiter's (with urethritis+conjunctivitis, *OHCM* p 407), ulcerative colitis, Crohn's, yersinia.

Ankylosing spondylitis (AS) *Prevalence:* 1 in 2000. Men are affected earlier ($\sigma/\eta \approx 6$ at 16yrs old and $\sigma/\eta \approx 2$ at 30yrs old). >95% are HLA-B27 +ve

Symptoms: The typical patient is a young man presenting with morning stiffness and progressive loss of spinal movement (spinal ankylosis) and sacroiliac joint involvement—with eventual kyphosis, neck hyperextension (question mark posture), and spinocranial ankylosis. *Other features:*

- Thoracic excursion↓
- Periostitis of calcaneum or ischial tuberosities
- Amyloidosis
- Chest pain
- Carditis; uveitis
- Hip/knee involvement
- Plantar fasciitis (p 642)
- Lung fibrosis (apex)

Radiology: 'Bamboo spine', vertebral squaring, erosions of apophyseal joints, obliteration of sacroiliac joints (sacroiliitis also occurs in: Reiter's; Crohn's; chronic polyarthritis). MRI may diagnose sacroiliitis earlier. *medline:*10752500

Blood: Normochromic anaemia; ESR↑. *Mortality:* 1.5-fold more than expected from amyloid or heart causes, but most never need hospital. *Rx:* Exercise, not rest, for backache and to optimize posture. NSAIDs may relieve pain and stiffness. Rarely, spinal osteotomy is useful. Tumour necrosis factor blockers (twice-weekly etanercept 25mg sc) can help active disease. *NEJM 2002 medline:*11986408

Gout When acute, there is severe joint pain, redness, and swelling, often of the metatarsophalangeal joint of the big toe (podagra). Attacks are due to hyperuricaemia (*OHCM* p 688) and deposition of sodium monourate crystals in joints—precipitated by trauma, surgery, starvation, infection, and diuretics. Urate deposits (tophi) form in avascular areas, eg pinna, tendons and joints (=chronic tophaceous gout). 'Secondary' causes of gout: polycythaemia, leukaemia, cytotoxics, renal failure. NB: in alcoholics, acute flare-ups may occur at lower plasma urate levels than in controls. *medline:*8035396

Diagnosis: Look for urate crystals in tissues & synovial fluid (serum urate not always↑). Microscopy of synovial fluid: negatively birefringent crystals; neutrophils (with ingested crystals). *Radiography:* Early, soft-tissue swelling. Later: 'punched out' lesions in juxta-articular bone. There is no sclerotic reaction; joint spaces are preserved until late. *Prevalence:* ~1/2–1%. $\sigma/\eta \approx 5:1$

Treating acute gout: Try an NSAID (eg naproxen 750mg stat, then 250mg/8h po pc). If contraindicated (eg peptic ulcer), give colchicine 1mg po initially, then 0.5mg/2–3h po until the pain goes or D&V occurs, or 6mg has been given

Prevention of attacks: Avoid prolonged fasts and purine-rich foods: offal; oily fish; roes; anchovies; mussels; crabs; prawns; shrimps; meat and yeast extracts; spinach; chocolate; caffeine, eg in cough-and-cold remedies. Avoid obesity and alcohol excess and very high-protein meals. No aspirin (it ↑ serum urate)

Consider reducing serum urate with long-term allopurinol, but not until 3 weeks after an attack. Allopurinol: 100mg/24h po pc, ↑ as needed to 600mg per 24h. Monitor serum urate levels. SE: rash, fever, WCC↓. If troublesome, substitute a uricosuric; get expert rheumatological help.

Pseudogout (calcium pyrophosphate arthropathy/chondrocalcinosis) *Risk ↑ if:*

- Old age
- Dehydration
- Hyperparathyroidism
- Myxoedema
- PO_4^{3-} ↓; Mg^{2+} ↓
- Osteoarthritis (OA)
- Haemochromatosis
- Acromegaly

Acute pseudogout is less severe and longer-lasting than gout, and affects different joints (mainly knee); chronic forms are like OA. Crystal deposits are punctate or linear, eg in the meniscus or parallel to the subchondral bone end plate. *medline:*7774886 Synovial fluid crystals are positively birefringent in polarized light. *Rx:* NSAIDs; if CIs, consider triamcinolone 60mg IM stat. *medline:*919552

*Controversial, but see *medline:* 11346223 & 10443078 & 8377534 & 1657004 & 11292305 & 627355

Reactive arthritis

This is an aseptic asymmetric mono- or oligoarthritis (ie just a few joints affected) developing ~1 week after infection elsewhere. 60–90% are HLA-B27 positive. Reiter's is a post-venereal or post-enteric example (OHCM p 426). Others: *Yersinia*, OHCM p 595 • *Chlamydia trachomatis*, p 512 • *Campylobacter* & *Chl. pneumoniae*, OHCM p 160 • *Ureaplasma*; HIV • *Salmonella/Shigella* • *Vibrio parahaemolyticus* • *Borrelia burgdorferi*, OHCM p 602 • *Clostridium dif.*

Polysaccharide antigen stimulation (causing IgA antibodies) may be important. **Treatment:** NSAIDs, physiotherapy, steroid joint injections. Antibiotics only help after yersinia or chlamydial infection. medline:10599344

Fibromyalgia (fibrositis)

Presentation: There is diffuse musculoskeletal (osteoarticular) pain of unknown cause, with morning stiffness, paraesthesiae, tender points over the body, skin-fold tenderness, and constitutional upset (sleep disturbance, fatigue, vertigo). It is a common 'diagnosis' made in rheumatology clinics in those who have pain with no obvious cause. But in the general population, most people with chronic widespread pain do not have high tender-point counts (below)—and most people with high tender-point counts do not have chronic widespread pain: so fibromyalgia may not be a distinct entity in the general population. Furthermore, it has been commented that the definition is circular. This may be true when a consultant says 'What's the cause of this man's pain?', and you reply 'Fibromyalgia'. What you should really reply is 'I do not know the cause, but the pattern of his illness suggests that its course will follow that typified by the family of patients bearing the label "fibromyalgia"'. medline:10199368 NB: beware diagnosing 'diffuse pain', unless all 4 quadrants of the body are affected, and there is axial pain.

Associations: Raynaud's, sicca & irritable bowel syndromes, anxiety/depression. **Modulating factors:** Temperature/humidity change, stress, fatigue.

Diagnosis: This is based on finding a number of separate characteristic tender points—when palpated with the thumb to give a pressure of about 4kg. It is usual to have ≥ 15 sites affected (left and right). The sites are:

Occiput at the semispinalis muscle insertion.

Low cervical at anterior aspect of intertransverse spaces at C5–7.

Trapezius at the mid point of the upper border.

Supraspinatus origins above the scapula spine near medial border.

Second rib surface just lateral to the second costochondral junction.

Lateral humerus near the epicondyles.

Gluteal at the upper outer quadrants.

Greater trochanter just posterior to the trochanteric prominence.

Knee at the medial fat pad proximal to the joint line.

Prognosis: The cause is unknown.

Treatment: is supportive. A randomized trial has shown benefit from electroacupuncture, but its use long term has not been assessed. medline:1477566

Other treatments tried: physiotherapy, NSAIDs. Antidepressants are efficacious in treating many of the symptoms of fibromyalgia. Whether this effect is independent of depression needs further study. medline:11029681

Osteoporosis

Osteoporosis implies reduced bone density. If trabecular bone is mostly affected, crush fractures of vertebrae are common; if cortical bone is mostly affected, fracture of a long bone is more likely, eg femoral neck: the big cause of death and orthopaedic expense, especially in older women.

Prevalence: 5%. $\text{♀}/\text{♂} \approx 4$ **Risk of future osteoporotic fracture** is increased if:

- | | | |
|---------------------------|---------------------|-----------------------------|
| • Slender or anorectic | • Early menopause | • Osteoporosis in family |
| • Smoker or alcoholic | • Cushing's disease | • Primary biliary cirrhosis |
| • Prolonged rest; old age | • Malabsorption | • Rheumatoid arthritis |
| • Hyperparathyroidism | • Thyrotoxicosis | • Hypogonadism |
| • >5mg/day prednisolone | • Myeloma | • Past low-trauma fracture |
| • Vertebral deformity | • Amenorrhoea | • Mastocytosis (p 602) |

Diagnosis: Once fractured, it is easy to look at a bone's image and say that osteoporosis is present. Without the 'retrospectroscope' this is harder. Serum Ca^{2+} , PO_4^{3-} and alkaline phosphatase are normal. Measurement of bone mass is via dual energy x-ray absorptiometry (DEXA). Screening with DEXA is probably not effective, and it is less effective than the taking into account of risk factors in the prediction of hip fracture. It provides little information about microstructure and elasticity—which are indirectly measured by ultrasound attenuation. So it is suggested, as the result of a large trial ($N=5662$ women with a mean age of 80 years) that if we combine DEXA with os calcis ultrasound we can identify groups with very high or very low risk of hip fracture. NB: biopsy specimens may be unrepresentative.

Prevention and management: In those who have had fractures recommend:

- Gentle exercise (including bone stress by impact) delays mineral loss.
- Continue using statins, if prescribed (may ↓ fracture risk by $\leq 60\%$).^{medline:11871921}
- If dietary Ca^{2+} ↓, give calcium with vit D (eg 1 or 2 sachets of Cacit D3[®] daily).
- HRT (p 18) delays perimenopausal bone loss. ▶ Avoid unopposed oestrogens (risks endometrial cancer). Raloxifene, a new SERM HRT, protects bones and reduces breast and endometrial cancer risk. See p 18. Trials of crystalline hydroxyapatite, fluoride, calcitonin, anabolic steroids, and vitamin D either show no additional benefit compared with HRT, or are seriously flawed
- In vertebral osteoporosis, consider long-term osteoclast inhibition with eg alendronic acid 70mg/week ½h ac (reduces risk of hip and wrist fracture too; SE: rashes, constipation; oesophagitis—so patients must wash down the tablet with plenty of water, and remain sitting up for 30min).

If on long-term steroids, bisphosphonates ↑ spinal density; calcitriol 6µg/day PO prevents bone loss and possibly fractures.^{medline:11846338}

Osteomalacia and **hyperparathyroidism** See OHCM p 702 & p 298.

Paget's disease of bone

There is ↑ bone turnover associated with abnormal hypernucleated osteoclasts causing remodelling, bone enlargement, and weakness. Rare in <40yrs; incidence ↑ with age. It is common in temperate climes, and in Anglo-Saxons. It may be asymptomatic or cause pain ± enlargement, eg of skull, femur, clavicle, tibia ('sabre' tibia) ± pathological fractures, nerve deafness (bone overgrowth) and high-output ccf. **Images:** Initially osteolytic; later mixed, then sclerotic, with expansion, coarse trabeculation, & deformity.

Tests: Plasma Ca^{2+} and PO_4^{3-} are normal; alk phos markedly raised.

Complications: Bone sarcoma occurs in ~10% of those affected for >10yr.

Treatment: Indications: • If symptomatic (eg bone pain) • To prevent fractures in long bones • In the axial skeleton, to prevent nerve damage. Use drugs which ↓ bone turnover, eg iv pamidronate (~30mg weekly for 6 weeks, perhaps repeated up to every 6 months) or oral bisphosphonate eg alendronic acid, above. Aim to normalize alk phos.

Densitometry MAY be indicated in those with:*

- Oestrogen lack which poses a substantial risk of osteoporosis:
 - Early menopause or premenopausal amenorrhoea.
 - Perimenopausal women if the result would change HRT plans.
- Vertebral osteopenia on plain radiographs.
- Some patients with vertebral deformity.
- Previous osteoporosis-related or 'low trauma' fracture(s).
- Monitoring response to osteoporosis therapy in some patients.
- Monitoring diseases associated with osteoporosis (opposite).
- Those taking long-term steroids.

Understanding DEXA scans: WHO osteoporosis criteria

Typical sites examined are the lumbar spine (preferably 3 vertebra) and hip. Bone mineral density (BMD, in grams/cm²) is compared with that of young healthy women (if it is a woman being tested). The 'T-score' relates to the number of standard deviations (SD) the BMD is from the average. If the T-score is:

- >0 BMD is better than the reference
- 0 to -1 BMD is in the top 84%: no evidence of osteoporosis
- 1 to -2.5 Osteopenia, with risk of later osteoporotic complication—so consider preventive measures (see opposite).
- 2.5 or worse BMD is in the bottom 2.5% of the population: osteoporosis is present—severe if there are one or more fragility fractures.

The T score predicts fracture risk: for every -1SD this risk doubles.

T-score = 0 suggests an average risk for a normal 40 year old.

T-score = -1 has twice the risk.

T-score = -2 has 4 times the risk.

T-score = -3 has 8 times the risk.

www.sw.org/ed/osteo/dexahow.htm

T-score decreases by -1 for *about* every 10% of bone lost (having 90% of young adult normal bone density yields a T-score of about -1.)

The Z-score is the standard deviations from normal for that patient's age.

If T normal and Z very abnormal, investigate the cause.

Osteoporosis in men* Consider densitometry in those with low-trauma fractures, or osteopenia on radiographs (or if on long-term steroids): If Z=0 to -1 (or T ≥ -2.5) offer lifestyle advice. If Z or T scores are more negative, do bone profile, FBC, TSH, protein electrophoresis & serum testosterone—and consider bisphosphonates (eg alendronic acid) ± anabolic steroids.* Recheck densitometry every 1–2 years.

Before acting on an abnormal DEXA scan consider other causes of low density: hyperthyroidism, hyperparathyroidism, myeloma, Cushing's, ankylosing spondylitis, rickets, premature menopause, vit D deficiency.

Other ways of measuring density Ultrasound measures at the heel.

DXA (single energy x-ray absorptiometry) measures at the wrist or heel

DPA (dual photon absorptiometry) measures at spine, hip, or total body

SPA (single photon absorptiometry) measures at the wrist

QCT (quantitative computed tomography) measures at spine or hip

RA (radiographic absorptiometry) uses an x-ray of the hand and a small metal wedge to calculate bone density.

PDXA (peripheral dual energy x-ray absorptiometry) measures at the wrist, heel, or finger.

http://imajinis.com/osteoporosis/osteo_diagnose.asp

Reducing pain in A&E

► All patients must be asked specifically if they require pain relief. Clinical assessment is expedited by relieving pain. Patients in pain must be given high priority and should be seen by a doctor as soon as possible. They should be reassured that their pain will be relieved promptly. No acute pain is uncontrollable. If systemic analgesia is required, select the analgesic most appropriate to the type of pain. In pleuritic or musculoskeletal pain, aspirin is often more efficacious than a narcotic. NB: There are NO contraindications to narcotic analgesia (other than well-defined allergy which is very rare). If in doubt—TREAT!

Adjuncts: Splint and elevate fractures and dislocations; RICE (p 663).

Local anaesthesia: Local infiltration of wounds with 0.5–1 % lidocaine; digital nerve block in finger injuries; femoral nerve block for fractured shaft of femur. *Non-narcotic systemic analgesia:* Paracetamol 1g/4h; aspirin 600mg ± codeine 15–30mg/3–4h. NB: avoid dextropropoxyphene (owing to dysphoria). NSAID: ibuprofen 400–800mg/8h po. Ketorolac is an injectable NSAID: 10–30mg IM or 10mg IV over 1min, then 5mg every 5min to a maximum of 30mg (if aged >65, max dose is 10mg). Tramadol is a synthetic opioid analgesic that has weak agonist activity at mu opiate receptors and inhibits serotonin and noradrenaline reuptake in the spinal cord. Tramadol dose: 50mg/4h po. *Gas:* Entonox® is a 50% mixture of nitrous oxide in O₂. It is self-administered by patients using a demand valve system and can provide a good level of analgesia for short procedures or when awaiting other forms of analgesia. *Ketamine* is a dissociative agent which can be used in subanaesthetic doses (ie: 0.25mg/kg IV) to provide analgesia. It should be used only under the authority of a consultant.

Narcotic analgesia must be given IV or IM (IM is too slow and unpredictable). *Morphine* dose: 50–100µg/kg, eg 2.5–7.5mg IV. Further increments should be titrated to the patient's response, initially at 5min intervals. Respiratory depression is unlikely with careful titration.

► There is no defined maximum dose. A typical total dose is 10–20mg. Don't give metoclopramide (≤10mg IV) routinely as vomiting is uncommon.

Fentanyl (1–2µg/kg, eg 50–100µg IV) is useful for short procedures requiring conscious sedation. (cardioversion, reduction of dislocated shoulder) especially in combination with midazolam.

Children with trauma needing pain relief White coats can be very frightening. Toys, friendliness, and a simple explanation of what is going to happen go a long way to avoid creating a lifelong fear of doctors.

- Do you need to suture a wound? Consider the alternatives (p 686).
- Do you need to inject local anaesthetic? Consider needle-free alternatives. It is rarely worth an injection for a single suture.
- If suturing or other painful procedures are unavoidable, consider sedation: morphine, fentanyl, or ketamine (p 792) are the agents of choice. Ketamine is a potent, short-acting analgesic at 3–10mg/kg. The injectable formulation can be used orally in a flavoured drink.

Explain gently to the child and the parents what you are going to do. Wrap the child in a blanket. Enlist enough nurses to hold the child and immobilize the part you are suturing. Warn the parents of possible distress. If they prefer not to be present, try to sit them out of earshot! Showing the child the result in a mirror, offering a big bandage, and giving an 'I was brave' badge can help. GA is rarely justified; if it is, the wound should probably be sutured by a plastic surgeon.

► Gunshot and other penetrating injuries

Murders by handguns: (Typical annual figures) Australia 13; UK 33; Sweden 6; USA 13,220. There are 4 cardinal rules in managing these injuries:

All but superficial wounds need exploration under GA to repair or exclude damage to deep structures (and for debridement). Get expert help as it is all too easy to do further damage.

Even minimal surface injury may hide much deep injury, depending on the weapon used, the mechanism of injury, and bullet speed.

Most victims are young and fit with good reserves. Therefore cardiorespiratory decompensation indicates severe injury.

If the weapon (eg knife) is still *in situ*, do not remove it until the patient is on the operating table—the danger is exsanguination.

Define and record each injury. Note that low velocity bullets leave dirty tracks, requiring exploration, excision, and *delayed* closure. High velocity weapons (eg military rifles) are extraordinarily destructive due to the temporary cavity caused by the missile. Ruthless and massive debridement is essential. Tetanus prophylaxis: p 686.

Chest injury Penetration of the chest may damage pleura, lung, great vessels, heart, mediastinum, diaphragm, and abdominal contents. The commonest injury is haemopneumothorax from damage to lung & chest wall. This requires a large (adult: 32 gauge) chest drain. Any deterioration or cardiac arrest demands prompt thoracotomy. Wounds of intercostal vessels, great vessels, or heart can cause massive haemorrhage: if drainage is initially >1500mL, or >300mL/h, thoracotomy is needed.

Sucking chest wounds must be closed at once, eg by Vaseline® gauze pads sealed on 3 sides only (acts as flutter valve)—complete the seal on chest drain insertion. ►► Relieve tension pneumothorax by needling the chest on the side of the suspected lesion *before* inserting a 32G chest drain (OHCM p 796) or doing radiographs (delay may be fatal).

Respiratory embarrassment due to pain, flail chest, or diaphragmatic injury requires intubation and ventilation. Insert a chest drain if there is any chance of a tear to lung, bronchus, or chest wall.

Cardiac tamponade 15% of deep chest injuries involve the heart.

Diagnosis: Beck's triad: rising venous pressure, falling BP, and a small, quiet heart (\pm pulsus paradoxus). NB: JVP may not be visible if there is hypovolaemia. ►► Pericardial aspiration is life-saving and diagnostic (and buys time before definitive anterolateral thoracotomy).

Procedure: Insert an 18G needle to the left of the xiphoid. Aim at the left shoulder, but with the needle angled downwards at 45° to the horizontal.

If these patients need: Crossmatching of ≥ 6 units of blood, THEN:

At least one large-bore IV cannula for vigorous fluid replacement.

Monitoring: vital signs; blood gases; CXR; ECG monitoring.

Intensive care with a chest drain immediately to hand, and facilities for immediate thoracotomy (eg if any deterioration, or a cardiac arrest).

Abdominal injuries All but the most superficial will need admission, and exploration, never just observation. See p 718.

Limb injury Nerves, tendons, and vessels are endangered, so examine limbs in good light, testing *pulses* (their presence doesn't exclude arterial injury), *sensation* and *sweating*. Any damage found will need formal surgical repair. Direct pressure will staunch bleeding. Do not use a tourniquet: may augment ischaemic damage, and you may forget to take it off.

The Glasgow Coma Scale (GCS)

This gives a reliable, objective way of recording the conscious state of a person. It can be used by medical and nursing staff for initial and continuing assessment. It has value in predicting ultimate outcome. 3 types of response are assessed:

- **Best motor response** This has 6 grades:

6 Carrying out request ('obeying command'): The patient does simple things you ask (beware of accepting a grasp reflex in this category).

5 Localizing response to pain: Put pressure on the patient's finger nail bed with a pencil then try supraorbital and sternal pressure: purposeful movements towards changing painful stimuli is a 'localizing' response.

4 Withdraws to pain: Pulls limb away from painful stimulus.

3 Flexor response to pain: Pressure on the nail bed causes abnormal flexion of limbs: decorticate posture.

2 Extensor posturing to pain: The stimulus causes limb extension (adduction, internal rotation of shoulder, pronation of forearm): decerebrate posture.

1 No response to pain.

NB: record the best response of any limb.

- **Best verbal response** This has 5 grades.

5 Oriented: The patient knows who he is, where he is and why, the year, season, and month.

4 Confused conversation: The patient responds to questions in a conversational manner but there is some disorientation and confusion.

3 Inappropriate speech: Random or exclamatory articulated speech, but no conversational exchange.

2 Incomprehensible speech: Moaning but no words.

1 None.

NB: record level of best speech.

- **Eye opening** This has 4 grades.

4 Spontaneous eye opening.

3 Eye opening in response to speech: Any speech, or shout, not necessarily request to open eyes.

2 Eye opening to response to pain: Pain to limbs as above.

1 No eye opening.

680

An overall score is made by summing the score in the 3 areas assessed, eg: no response to pain+no verbalization+no eye opening = 3. Severe injury, GCS ≤8; moderate injury, GCS 9–12; minor injury GCS 13–15.

Note: an abbreviated coma scale, **AVPU**, is sometimes used in the initial assessment ('primary survey') of the critically ill

● **A** = **alert**

● **V** = responds to **vocal** stimuli

● **P** = responds to **pain**

● **U** = **unresponsive**

►► Drowning

Worldwide, half a million drowning deaths occur yearly: most are children.¹ Boys are at especial risk. Drowning is second only to road crashes as the leading cause of death in those aged 1–40yrs.

Most drownings occur in drunk adults, or in children who are poorly supervised near water. It is quite common for a toddler to bath with a slightly older sibling 'in charge'—the mother thinking that as the child can sit up, he can also save himself from drowning should he topple over.

In communities wealthy enough to have many private swimming pools there are summertime spates of drownings. An adult is temporarily distracted and leaves the garden, and, on return, finds a corpse in the pool.

When such a child is brought to A&E the chief concerns are:

1 Cardiac arrest **2** Hypothermia **3** Acidaemia **4** Pulmonary oedema.

Management focuses on **airway**, **breathing** and **circulation**:

- Immobilize the cervical spine as you open the airway—there may have been trauma here.
- Clear vomit from mouth. Vomiting is common (much water is ingested).
- If pulseless, start cardiopulmonary resuscitation.
- Expect BP to drop after leaving the water, and be sure to transport in the horizontal position, and keep cardiovascular instability to a minimum.²
- Intubate (pre-oxygenate with 100% O₂; if hypothermic, this can induce vf). Use 100% O₂—water in alveoli dilutes surfactant, causing atelectasis.
- Do Glasgow Coma Scale, p 680; if <6 prognosis is poor.
- Monitor ECG. Defibrillate if in vf (p 311). For the first shock, give ~2 joules/kg body weight (~20J at 9 months, 40J at 4yrs, 60J at 8yrs, and 80J at 12yrs). NB: defibrillation often fails until core T° >30°C. Various heroic measures have been tried without necessarily affecting outcome³—eg rapid core rewarming, eg by warm-water immersion, warm iv fluids at 40°C, breathing of heated humidified gas,⁴ peritoneal dialysis, thoracotomy for heart irrigation, or cardiac bypass.
- Monitor core T° & CNS signs. Cerebral oedema may occur suddenly in first 24h: no evidence supports use of dexamethasone or barbiturate coma.
- If T° <35°C, monitor rectal T° often and rewarm using high ambient T°. Circulatory support may be needed to counter rewarming vasodilatation. Measuring left atrial pressure helps guide iv fluid: too much will ↑ICP.
- Pass a nasogastric tube to relieve gastric dilatation.
- U&E, blood gases, and Hb. Get expert advice on the problem of acidosis.

Effects of asphyxia and fresh water overload The child usually swallows large amounts of water before final aspiration. This leads to gastric dilatation, vomiting, and further aspiration.⁵ Acute respiratory distress syndrome can occur, leading to the major villain, hypoxaemia—hence cerebral oedema and ↑ICP. To combat these, steroids and antibiotics are sometimes given, but without proof of efficacy. As the raised intracranial pressure (ICP) is due to cell death (cytotoxic oedema), steroids do not lower ICP (unlike the vasogenic oedema seen with space-occupying lesions).

The diving reflex Children retain this useful adaptation to our earlier aquatic way of life. As cold water hits the face, the pulse slows and blood is diverted from limbs and muscles to vital areas (brain, kidney). Cold further reduces the metabolic rate. This is the physiological explanation for remarkable stories of recovery from prolonged (eg 20min) total immersion. It is also the reason why resuscitation of a seemingly dead child must continue for 40min before death is declared.

Preventing death by drowning

In richer parts of the world, drownings in garden ponds are getting commoner, while those in natural freshwater sites are declining. Toddlers have the highest drowning rates—so prevention must target toddlers:¹

- Constant supervision of infants in baths by adults. Even brief moments away, eg to answer the door-bell, can be disastrous.

- Isolation fencing that surrounds the pool, separating it from the home.

- Not swimming alone or in remote, unguarded sites.

- Training of the public in basic life support. If given promptly, neurological outcomes are better.^{6,7}

Brenner 2002 *BMJ* **i** 1050 <http://bmj.com/cgi/content/full/324/7345/1049>

Stoneham 1995 *Lancet* **345** 1048 **3** L Spack 1997 *Pediatr Emerg Care* **13** 98

Maier 1995 *Lancet* **345** 1048

5 J Orlowski 2001 *Pediatr Clin North Am* **48** 627 [11411297]

Kyriacou 1994 *Pediatrics* **94** 137

7 I Pless 1997 *Inj Prev* **3** 237 [9493614]

►► Major injury, including blunt chest injury

On news of major trauma, summon experienced help (eg senior traumatologist). Remember **ABC** (a irway, b breathing, c irculation): **OPPOSITE**. If breathing spontaneously, give *all* patients O_2 at 15L/min through tight-fitting mask with reservoir. *Quality of breathing*: stridor (\pm voice change) indicates a sternoclavicular fracture/posterior dislocation—get expert help at once as intubation may be difficult owing to tracheal compression, until the dislocation is reduced—by extending the shoulders, and grasping the clavicle with a clamp (eg towel clip) and manually reducing the fracture. Cover & seal open chest wounds on 3 sides only. ►► *Is there a tension pneumothorax?* Urgent treatment: chest needling on the side with \downarrow breath sounds; other signs are:

- Air hunger + respiratory distress – Pulse \uparrow ; BP \downarrow – Tracheal deviation
- Breath sounds \downarrow on one side – Cyanosis – Neck vein distension

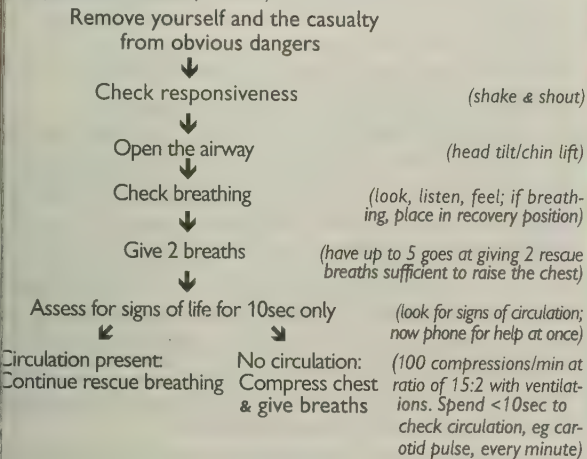
Relieve by inserting a needle (2nd intercostal space, mid-clavicular line).

- Apply pressure and elevation to any actively bleeding part.
- Crossmatch; glucose stix; 2 wide-bore saline ivs (femoral catheter or saphenous cutdown anterior to medial malleolus if no iv access). medline:9420119
- Take systolic BP. If <90 mmHg and blood loss is the probable cause, give colloid (Haemaccel[®]) fast iv until BP \uparrow , pulse \downarrow , urine flows (>30 mL/h; catheterize at leisure—exclude urethral injury first) and crossmatched blood arrives (► ask for group-compatible blood—only takes 5–10min).
- Remove clothes (large scissors). *What are the injuries?* Do circumferential burns need escharotomy to \downarrow laryngeal pressure? Any surgical emphysema (chest drain needed?). Is there a flail chest (a segment has no bony continuity with thorax, with paradoxical respiration)? The main problem is the injured lung. This is sensitive to over and under resuscitation; intubate & ventilate.
- Assume spinal instability. Before radiology, keep immobile (sandbags, collar, tape; CT is the best method if high/moderate risk for cervical fracture). medline:10405730
- Do level-of-consciousness (AVPU, p 680) + ECG (ST & conduction problems \approx myocardial damage: extent is revealed via direct inspection by a thoracic surgeon).
- Falling BP + rising JVP and a quiet heart = cardiac tamponade: ► see p 679.
- Examine all peripheral pulses. 30% of fractures are missed in resus rooms. Image cervical spine (p 726) + pelvis. Skull radiographs waste time. If intraperitoneal bleeding is suspected, see p 718. Do blood gases & cxx; implications:
 - If free air, needle the chest (or formal chest drain) for pneumothorax.
 - If persistent large pneumothorax after chest drain suspect bronchial tear.
 - # of ribs 1–3 \approx ?airway/big vessel injury* – Ribs 9–12 \approx ?abdominal trauma
 - # of >2 ribs in 2 places \approx ?flail chest – cxx fluid level \approx haemothorax*
 - Bowel gas in chest \approx ?diaphragm injury – Diaphragm contour \downarrow \approx ?rupture
 - Coils of NGT in chest \approx ?diaphragm injury – Liver raised \approx ?diaphragm injury
 - # of sternum \approx ?myocardial contusion – Mediastinal air \approx lung barotrauma
 - Respiratory distress + cxx OK \approx ?aspiration – # of scapula \approx ?airway injury
 - Wide mediastinum \approx ?aortic rupture* – Tracheal deviation \approx aortic injury*
 - Deviation of oesophagus \approx ?aortic rupture* – No aortic knob \approx ?ruptured aorta*
- Chart pulse, BP + pupil size every few mins. If pupils unequal, summon neurological help; give 20% mannitol 1g/kg (5mL/kg) iv (\uparrow cerebral blood flow).
- Head-to-toe exam ('secondary survey'). Glasgow Coma Scale (p 680).
- If fits occur give 10mg diazepam iv followed by 40mg in 500mL 5% dextrose infused at ~ 100 mL/h if they continue (children: 0.3mg/kg iv, then 50 μ g/kg/h ivi)—beware apnoea.
- In near-drowning or smoke inhalation, consider early high-dose methylprednisolone (30mg/kg iv); also in lung contusion (ventilate if $S_{A_{O_2}} < 90\%$).
- Give tetanus toxoid booster \pm human anti-tetanus immunoglobulin (p 686).
- Get experienced help in prioritizing treatment for each specific injury.

Have a low threshold for contrast-enhanced aorta CTs, especially if deceleration injury: ATLS 6e, 135

► Adult basic life-support algorithm

This algorithm assumes that only one rescuer is present, with no equipment. (If a defibrillator is to hand, get a rhythm readout, and defibrillate, as opposite, as soon as possible.)



Managing the airway

- You open the airway by tilting the head and lifting the chin—but only do this if there is no question of spinal trauma.
- Use a close-fitting mask if available, held in place by thumbs pressing downwards either side of the mouth-piece; palms against cheeks.

Chest compressions

- Cardiopulmonary resuscitation (CPR) involves compressive force over the lower sternum with the heel of the hands placed one on top of the other, directing the weight of your body through your vertical, straight, arms.
- Depth of compression: ~4cm.
- Rate of compressions: 100/min.

appears as an opaque hemithorax if the CXR is supine.

See www.resus.org.uk/pages/blsalgo.pdf Definitive control rather than normalization of plasma volume is the aim in bleeding^{ATLS} For practising dynamic scenarios with shifting scenarios see www.trauma.org/resus/moulage/moulage.html

The stitching of wounds

- 1 Infiltrate skin to be sutured with 1% lidocaine (<3mg/kg; SE: seizures); wait 3min for it to work. ► **Avoid adrenaline (epinephrine) when anaesthetizing digits.** Remove foreign bodies and all necrotic material. Clean with 1% cetrimide. Control bleeding by pressure and elevation of the part. Deep absorbable stitches (eg Dexon®, Vicryl®, catgut) will aid apposition of skin edges (avoid in the hand, as may disrupt nerves).
- 2 If much tension is needed, consider tension sutures or skin grafts.
- 3 Handle the tissues gently to prevent further injury.
- 4 Use interrupted stitches for closure. Start from the middle, or with any jagged edges. This aids accurate apposition.
- 5 Avoid eversion or inversion of the stitched skin edges.
- 6 Examples of which stitches to use: *Adult's face:* 5-0 monofilament nylon (remove after 5 days). *Child's face:* 6-0 monofilament nylon (remove after 3–5 days). *Other parts of children:* 5-0 catgut. The deep part will be absorbed and the top part will slough off after 10–14 days. *Adult leg:* 3-0 nylon; remove after 2 weeks. *Shin skin flaps* (after a fall on a hard object) may have a poor blood supply, especially if the length of the flap is more than twice as long as its breadth, and healing will be better with thin strips of special adhesive tape (eg Steristrip®), rather than stitches. Exact approximation of the edges is likely to be impossible, but skin grafts are rarely needed. *Adult arm, trunk, abdomen:* 3-0 silk (or equivalent). Remove after ~9 days. *Scalp:* 3-0 silk. Remove after 5 days.
- 7 Learn the technique from an experienced professional (eg staff nurse). Practise suturing a slashed banana skin or pig's trotter in a sterile field.
- 8 Dress with sterile tulle to prevent further injury.

Alternatives to suturing Suturing requires anaesthesia and leaves scars. *Skin closure strips (Steristrips®)* are useful for cuts on non-hairy skin. Leave space between strips for ooze. They can also temporarily appose edges of larger wounds while you suture. Infection is less of a risk than with stitches, particularly for animal bites. *Acrylic tissue adhesive (Histoacryl®)* is quick, minimizes scarring and needs no follow-up. Wear gloves to stop your flesh sticking to the patient. Dry the wound, and align edges carefully: you cannot correct the position once the sides have touched. Apply glue thinly (use a capillary tube), and press the edges together for 30sec.

Preventing tetanus Active immunization with tetanus toxoid is given as part of routine vaccination at 2, 3, and 4 months of age (as DTP, p 209). A 4th dose is given >3yrs later, and a 5th dose before leaving school (as 'Td', see below). Once given 5 injections at the allotted intervals, no further toxoid is generally needed, and risks side-effects, and ↓immunity by overstimulation. If travel to areas without access to immunoglobulin, further boosters may be needed if >10yrs since the last dose.

Primary adult immunization. 0.5mL of 'combined tetanus & low-dose diphtheria vaccine' (Td) IM repeated × 2 at monthly intervals with boosters at 10 & 20yrs.

Wounds: Any cut merits an extra dose of 0.5mL toxoid (Td) IM, unless already fully immune (5 doses as above). The non-immune will need 2 further injections (0.5mL IM) at monthly intervals.

Human tetanus immunoglobulin is for the partially or non-immune (defined above) with infected, devitalized, soil-contaminated, or puncture wounds. Give 250–500 units IM, using a separate syringe and site to the Td. Manure-contaminated wounds need treating this way even if fully immune.

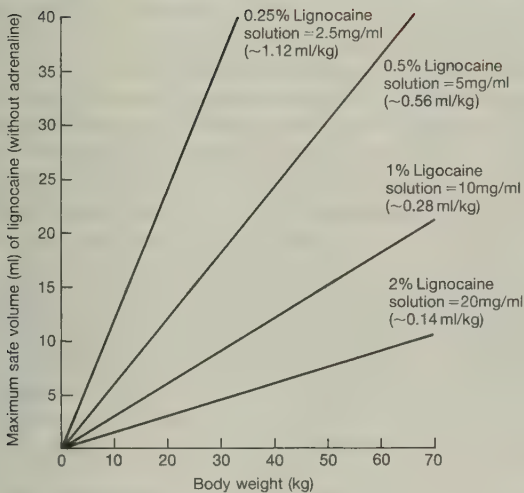
► If the immune status is unknown, assume he/she is non-immune.

Routine infant immunization started in 1961, so many adults are at risk.

► Hygiene education and wound debridement are of vital importance.

Maximum doses of lidocaine: a rough guide¹

0.25%	1.12mL/kg
0.5%	0.56mL/kg
1%	0.28mL/kg
2%	0.14mL/kg



Maximum safe volume of plain lidocaine (=lignocaine) according to body weight. (Maximum safe dose of lidocaine—without epinephrine (=adrenaline) 3mg/kg, with adrenaline 7mg/kg; uses of different solutions—0.25–0.5% for infiltration and intravenous regional anaesthesia, 1% for nerve blocks, epidural anaesthesia, and intravenous regional anaesthesia, 2% for nerve blocks.)

►► Major burns (>20% of body area)^{ATLS}

Resuscitate and transfer quickly to a burns unit. Assess site, size, and depth of the burn. Refer full thickness burns >5% of body area or full/partial thickness burns involving face, hands, eyes, or perineum for specialist care.

Assessment The size of the burn must be estimated to calculate fluid requirements. Ignore erythema. Use a Lund & Browder chart (special charts for children and neonates that take account of differences in body proportion with age) or the 'rule of nines' in adults: medline:8726264

Arm (all over) 9%	Front 18%	Head (all over) 9%	Palm 1%
Leg (all over) 18%	Back 18%	Genitals 1%	

(Although for ATLS purposes, the palm is 1%, strictly this includes the palmar surface of the fingers too.)

Estimating burn thickness can be difficult: superficial dermal is painful, red, and blanches readily; full thickness is painless and white/grey with fixed staining. Do not include simple erythema as a burn.

Resuscitation *Airway:* Beware airway obstruction if hot gases inhaled—suspect if history of enclosed space, singed nasal hairs, soot in mouth, or changed or hoarse voice. Involve anaesthetists early (eg early intubation).

Breathing: Give 100% O₂ if you suspect carbon monoxide poisoning. NB: he may have cherry-red skin. The t_{1/2} carboxyhaemoglobin (COHb) is long; consider hyperbaric oxygen if pregnant or CNS signs, or >20% COHb. SpO₂ measured by pulse oximeter can be unreliable. Consider trunk escharotomy if thoracic burns impair chest excursion (p 689).

Circulation: >10% burns in a child and >15% burns in adults require iv fluids. Put up 2 large-bore (14–16G) lines. Don't worry if these go through burned skin; intraosseous access is valuable in infants (p 309). Secure them well: they are lifelines. Use a 'Burns Calculator' flow chart or a formula:

Parkland formula (most commonly used in Burns units) $4 \times \text{weight (kg)} \times \% \text{burn} = \text{mL crystalloid usually Hartmann's solution in 24h, half given in first 8h.}$

Muir and Barclay formula $[\text{weight (kg)} \times \% \text{burn}]/2 = \text{mL colloid (eg Haemaccel®)} \text{ per unit time. Time periods are 4h, 4h, 4h, 6h, 6h, 12h.}$

Either formula is acceptable but you must use the appropriate fluid, ie Crystalloid not colloid for Parklands. Replace fluid from the time of burn, not from the time first seen in hospital and don't forget maintenance fluids. You must also give 1.5–2.0mL/kg/h 5% dextrose if Muir and Barclay formula is used. Formulae are only guides: adjust iv rate according to clinical response and urine output (aim for >30mL/h in adults; >1mL/kg/h in children; and more in both cases with inhalation injury). Monitor T° (core & surface) and catheterize. Ensure tetanus immunity. Give 50mL whole blood for every 1% of full-thickness burn, half in the second 4h of iv therapy and half after 24h.

688

Transfer dressing Don't apply cold water to extensive burns: this may intensify shock, and cooling of a burn is only effective in the first minutes of injury. Avoid the use of specific burns dressings, leave this to the burns unit unless previously agreed (delayed presentation/contaminated burn) as some creams may hinder assessment. After toileting, cover extensive partial thickness burns simply with clingfilm prior to transfer. Keep the patient warm. Use morphine in 1–2mg aliquots iv; adequate dosing is important.

Definitive dressings/treatment Superficial burns are dressed with EZderm (porcine) or mepitel (silicone). Mixed depth burns are commonly dressed with silver sulfadiazine (flamazine) but requires changes every 1–2 days. (Contraindicated in pregnant/ lactating women and neonates less than 2 months). Hands may be covered in silver sulfadiazine inside a plastic bag/ loose dressing. Elevation is vital. Major full thickness burns benefit from ETE (early tangential excision) and ssg (split skin grafting). Burns wounds are a

major source of inflammatory mediators causing SIRS and they form an ideal medium and entry point for bacteria. Early excision is life saving.

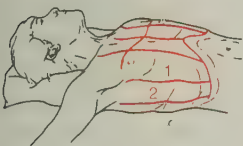
Paediatric burns $\frac{2}{3}$ are scalds. Because of a larger surface area : volume ratio and fewer thermoregulatory compensatory mechanisms, there is greater tendency to hypothermia. Because of different body proportions the 'rule of nines' has to be altered. Be alert to non-accidental injury.

Smoke inhalation Initially, there is laryngospasm that leads to hypoxia and straining (causing typical petechiae), then hypoxic cord relaxation occurs and at this point true inhalation injury occurs. Free radicals, acids, cyanide poisoning, and carbon monoxide poisoning accompany thermal injury. Cyanide poisoning is common from smouldering plastics. It binds reversibly with ferric ions in enzymes—uncoupling oxidative phosphorylation and causing dizziness, headaches, and seizures. Increased ventilation by peripheral chemoreceptor stimulation makes toxicity worse. Tachycardia and dyspnoea soon give way to bradycardia and apnoea. Carbon monoxide poisoning is a relatively late feature of inhalation injury as the fire runs out of oxygen; the level of blood carboxyhaemoglobin does not correlate well with the severity of inhalation injury.

100% oxygen is given to elute cyanide and carbon monoxide. Involve anaesthetists and ICU early: early ventilation may be indicated, and consider bronchoscopy with repeated bicarbonate lavage. In cyanide poisoning, get expert help. There is no single treatment regimen that is best for all situations. Individual risk factors for antidote toxicity, the nature of the exposure, and its likely severity, and the proximity to hospital facilities all need considering. Mild poisoning may be treated by rest, O_2 , and amyl nitrite 0.2–0.4mL via an Ambu[®] bag. Intravenous antidotes are indicated for moderate poisoning. Where the diagnosis is uncertain, sodium thiosulfate may be first choice. With severe poisoning, an additional agent is required. Hydroxocobalamin may be best. Less safe agents: sodium nitrite solution, dimethylaminophenol, and sodium thiosulfate.

medline:10024740

Escharotomy For chest escharotomy make incision 1 first. If further expansion is needed make the others. Make incisions either side of trunk/limb or digit (one side only shown below). If it causes pain, burns are not full thickness and escharotomy is probably not needed. Bleeding may prompt covering with sterile dressings \pm transfusion. www.txt.co.uk/fu1.cfm?0689



Fractures

Fractures may be *transverse*, *oblique*, or *spiral*. They are *comminuted* if there are >2 fragments. If broken ends protrude through the skin or into a body cavity, the fracture is *open* (an emergency). Young soft bones may bend and partially break (greenstick fractures). Compression forces yield *impacted* or *crush* fractures (in cancellous bone), and traction force causes tendons or ligaments to detach a piece of bone (*avulsion*). A *complicated* fracture causes significant damage to vessels, nerves, or nearby organs. Separation of the parts is defined in terms of side-to-side *displacement*, *rotation*, and *angulation*. There may be dislocation or subluxation (partial loss of contact of joint surfaces). Ask about the degree of force which produced the injury. If it seems trivial, the bone may be diseased (*pathological* fracture, eg in secondary deposits, osteoporosis, Paget's disease—p 676).

Clinical features Pain; loss of function; tenderness; deformity; swelling; crepitus (grating of the broken ends—be gentle!).

Radiographs • How many fractures are there? Can you see the whole bone?

- Alignment (displacement, angulation, rotation)
- Are the bones normal? Look for trapped air, bone pathology and any foreign body. For further help, see *Medical Imaging* (oup, page 87).
- Are more radiographic images needed (ivu, ct, arteriogram)?

Salter and Harris classification of epiphyseal injury (See diagram)

- I Seen in babies or pathological conditions (eg scurvy).
- II The commonest.
- III There is a displaced fragment.
- IV Union across the growth plate may interfere with bone growth.
- V Compression of the epiphysis causes deformity and stunting.

Treatment • Correct shock (eg colloid iv, 500mL/30min, until systolic BP >100mmHg); give blood if >1–1.5 litres lost, or continued bleeding. Monitor BP/15min, urine output (\pm cvp). Stop bleeding (may need open surgery).

- Relieve pain (eg morphine 10mg/4h im or 1–2mg aliquots iv if shocked).
- Sterile cover (eg Betadine®-soaked) over protruding bones and torn skin.
- Prevent infection: eg cloxacillin 500mg im/iv + benzylpenicillin 600mg iv/im; tetanus toxoid (0.5mL im) if needed.

Ways of managing fractures: Immobilize and reduce the part (if needed, p 692), eg using a plaster of Paris cast. The problem with this method is that 'fracture disease' follows immobilization—muscle atrophy, stiff joints, and osteoporosis—so anything speeding return of function may be beneficial.* If possible, fractures involving joint articulations should be treated by open reduction, accurate reconstruction of the joint surfaces, and fixation, such that immediate movement can occur. Otherwise, secondary osteoarthritis is inevitable. Prompt *internal fixation*: eg with plates (~10 screw-holes), nails (Küntscher) or wires (Kirschner)—of all fractures in those with poly-trauma leads to large reductions in serious complications (fat embolism, acute respiratory distress syndrome), as well as lessening the time during which mechanical ventilation is needed.

medline:10721407

External fixation uses screws into bone, a bar, and a means (cement or clamps) of attaching the bar to the screws to align and engage the fractured cortices. The screws may be driven through the bone and out of the far side of the limb, so that a second bar can add stability. Further rigidity is afforded by another set of screws at 90° to the first set. Because the intervention is away from the field of injury, this method is very useful when there are burns, loss of skin and bone, or an open fracture.

*J Busse 2002 *CMAJ* 166 437 meta-anal (@11873920); it's nonstandard; healing may be speeded by ~64days.

Fixation options

Kirschner wires

Staples

Osteosynthesis lag screws

Absorbable pins

Cannulated screws

Self-tapping screws

Herbert headless screws

T-screws

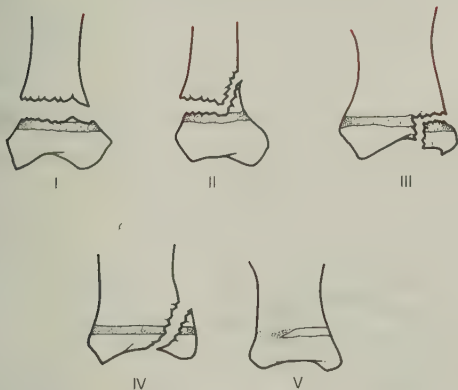
medline:11098739

www.osteomedcorp.com/articles/

Screw insertion to unite the fragments typically begins with temporary wire or bone clamp fixation. A pilot hole is then drilled into bone, which must be substantial enough to support the screw head. Both near and far cortices are drilled with the pilot drill. Countersinking decreases incidence of stress risers and microfractures of the bone as the head of the screw compresses into the near cortex. The pilot hole is measured to aid selection of the best length of screw. Distal threads have a smaller diameter than proximal threads to give thread cutting action for self-tapping screws. Have eg 2 threads of the screw exiting the far cortex to assure that the largest threads of the screw purchase the far cortex. (Add eg 2mm to the pilot measurement.)

Lag screw technique: An oversized countersunk hole is drilled through the near cortex. The distal portion of the hole is drilled with a diameter corresponding to the core diameter of the screw. As the screw tightens its distal end engages the threaded hole of the distal fragment. The undersurface of the screw head contacts the countersink hole and pulls the distal fragment against the proximal fragment causing compression. A lag effect can only occur if the screw can pass freely through the gliding hole. Lag screws are most appropriate for oblique fractures.

www.bcm.tmc.edu/oto/grand/42993.html



Fracture reduction and traction

A displaced fragment needs realignment (reduction) unless function & appearance of the limb are satisfactory. For example, a small displacement acceptable in a fracture of the femoral shaft might not be acceptable in the radius, where it could cause difficulty in supination. Reduction also allows freeing of any structures trapped between bone ends. Accurate reduction aids revascularization (vital in subcapital fractures of the femur) and prevents later degenerative change if fractures involve joints. Internal and external fixation has removed the need for much traction in adults, but it is still used for children

Methods of reduction *Manipulation* under anaesthesia (most cases).

Traction may be used for subcapital femur fractures or spinal injury.

Open reduction aids accuracy, eg before internal fixation (p 690).

The next problem is to hold the reduction in place (using traction or fixation) during healing—which may take from 2 weeks (babies) to 12 weeks (a long bone in an adult).

Skin traction uses adhesive strapping to attach the load to the limb. The problems are that the load cannot be very great, and that sensitivity to the adhesive may develop.

Bone traction Using a pin through bone, bigger forces can be employed (eg Steinmann pin, Gardner-Wells, or Crutchfield skull calipers).

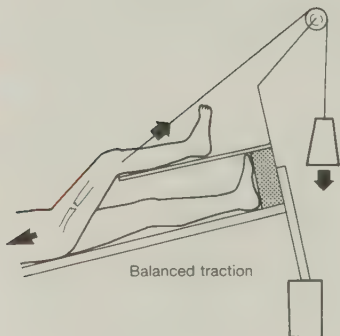
Fixed traction The Thomas' splint.

Balanced traction (below) The weight of the patient balances against the load.

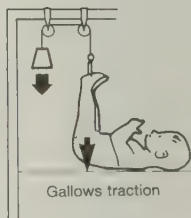
Gallows traction is suitable for children up to 2 years of age. The buttocks rise just above the bed.



Thomas' splint



Balanced traction



Gallows traction

Complications from fractures

Healing Haematoma → Vascular granulation tissue → Subperiosteal osteoblast stimulation → Bone matrix → Endochondral ossification → Deformable woven bone (callus) → Lamellar bone → Fracture union.

Complications Immediate:

Internal or external bleeding
Organ injury, eg brain, lung
Nerve or skin injury
Vessel injury (limb ischaemia)

Later local:

Skin necrosis
Gangrene
Pressure sores
Infection^{1,2}

General:

Venous thrombi
Pulmonary emboli
Pneumonia
Renal stones

Fat embolism typically occurs on day 3–10. Signs: confusion, dyspnoea, $P_aO_2 \downarrow$, fits, coma, $T^{\circ} \uparrow$, petechial rash, CXR/CT: small subpleural nodular opacities. medline:10634667 Management: take to ITU, get expert help, correct shock (monitor CVP & urine output). Treat respiratory failure (OHCM p 178). Aim for early mobilization. Early fixation of fractures is preventive.

Crush syndromes & compartment syndromes are a cause of ischaemia; also fluid loss, DIC, and myoglobin release cause tubular necrosis (dialysis may be needed). Correct hypovolaemia vigorously. Watch urine output and plasma K^+ . A limb's own dead weight when immobile or in coma, may cause crush injury/compartment syndrome—a vicious cycle of pressure → vascular occlusion → hypoxia → necrosis → pressure ↑. Look for tell-tale signs of redness, mottling, blisters, swelling, and pain on passive muscle stretching. ▶ Compartment syndrome is both a limb- and life-threatening emergency. Avoiding delay requires vigilance and, if necessary, intracompartmental pressure measurement—but reference ranges for intervention are controversial. Prompt fasciotomy prevents contractures (p 616) and amputations. meta-analysis medline:11952578

Late complications

- Wound sepsis (early wound cleaning is vital). Infection also complicates ~8% of closed fractures if internal fixation is used.^{3,4}
- Failure of fixation (eg plates or nails break, or dislodge).
- Joint stiffness, contractures, malalignment (malunion).
- Algodystrophy (see OPPOSITE): joint stiffness and patchy osteoporosis.
- Psychological problems in mobilizing ('compensation neurosis').
- Non-union implies mobility at the fracture site at 6 months (provided you are satisfied that all healing potential has been used up). Mobility at 3 months is *delayed* union. Non-union is commoner in cortical bone than in cancellous bone, where there is often impaction. **Factors delaying union:**
 - 1 A fracture in a bone which has finished growing.
 - 2 Poor blood supply (eg lower tibia) or avascular fragment (eg scaphoid).
 - 3 Comminuted/infected fractures, eg after open reduction & internal fixation.
 - 4 Generalized disease (eg malignancy, infection).
 - 5 Distraction of bone ends by muscle; open reduction ± internal fixation prevents this. NB: osteoporosis ± old age don't necessarily delay union.

Management of non-union Rigid immobilization helps (eg internal fixation). If there is no callus, a cortical bone graft may help by providing both fixation and an osteogenic stimulus. It may be inlaid, or screwed on top of the bone ends. If realignment is not necessary a cancellous graft (using marrow-rich chips) may be used (± internal fixation). If infection is present, dead bone must be removed. Bone chips then fill in the space, which is held open by external fixation. Note: inducing an alternating electromagnetic field across the fracture appears to give encouraging results.

Algodystrophy

Synonyms

Croft's atrophy/osteodystrophy
reflex sympathetic dystrophy; sympathetically maintained pain syndrome
post-traumatic sympathetic atrophy;
shoulder-hand syndrome
minor causalgia (causalgia means burning pain)
post-traumatic painful osteoporosis
Complex regional pain syndrome type I

Definitions 'A complex disorder of pain, sensory abnormalities, abnormal blood flow, sweating, and trophic changes in superficial or deep tissues.' The central event may be loss of vascular tone or supersensitivity to sympathetic neurotransmitters. The 1994 systematic criteria for definitive diagnosis are given below*, but are of uncertain validity.

2002 meta-analysis
medline:11865127

Presentation This may follow weeks or months after an insult—which may be minor trauma, a fracture, zoster, or myocardial infarction. Lancing pain (which may have a trigger point) in a limb accompanies vasomotor instability. The limb may be cold and cyanosed, or hot and sweating (locally). Temperature sensitivity may be heightened. The skin of the affected part may be oedematous—and later becomes shiny and atrophic. Hyperreflexia, clonus, and contractures may occur.

Note that it is not the traumatized area where the symptoms occur: rather, it is some neighbouring area.

There are no systemic signs (no fever, tachycardia, or lymphadenopathy). Anxious, neurotic personalities are particularly affected—perhaps because of poor mobilization following the initial insult.

Tests Patchy osteoporosis on radiography, but no joint space narrowing (thinning of cartilage). Bone scintigraphy shows characteristic uniform uptake.

Treatment Refer to a pain clinic. Standard painkillers often have little effect. Consider physiotherapy and NSAIDs. Calcitonin and postganglionic sympathetic blockade (guanethidine or bretylium) have advocates. Ultimately, the condition is self-limiting.

Continuing pain which is disproportionate to any inciting event with reports of at least one **symptom** in each of these 4 categories: Sensory; vasomotor; sudomotor/oedema; motor/trophic—and at least one **sign** in 2 or more of the above categories. www.stoppain.org/for_professionals/archive/pain_jul99.html#assessment

Trauma to the arm

Fracture of the clavicle This is common in the young, eg from a fall on an outstretched arm (which in the elderly would be more likely to cause a Colles' fracture). A broad sling support for 3 weeks is usually all that is needed. Fractures at the lateral end may need internal fixation.

Scapula and acromion fractures and acromioclavicular dislocation rarely require anything more than sling support. Mobilize early.

Fracture of the humeral neck Often these are impacted and stable; if there is displacement there is risk to the brachial plexus and axillary artery. Manipulation or open reduction may be needed if there is displacement of the upper humeral epiphysis (in children) or if there is co-existing shoulder dislocation. Fractures of the head itself may result in 2–4 fragments. If ≥ 3 , internal fixation is likely to be needed.

Fracture of the humerus shaft This is often caused by a fall on an outstretched arm. Marked displacement often makes the diagnosis easy. Radial nerve injury may cause wrist-drop. Splinting with metal gutter-shaped splints and support from the wrist to the neck (collar and cuff sling) usually gives satisfactory reduction. Immobilize for 8–12 weeks.

Anterior shoulder dislocation may follow a fall on an arm or shoulder. *Signs:* Loss of shoulder contour (flattening of deltoid), an anterior bulge from the head of the humerus, which may also be palpated in the axilla. Check pulses and nerves (including the axillary nerve supplying sensation over lower deltoid area) pre- and post-op. Before reduction, do radiography (is there a fracture too?)—unless this is a case of recurrent dislocation, when just a post-reduction image may be needed.

Relieve pain (eg parenteral opioid, and Entonox[®] through the procedure). Methods of reduction:

Treatment by simple reduction: Apply longitudinal traction to the arm in abduction, and replace the head of the humerus by gentle pressure.

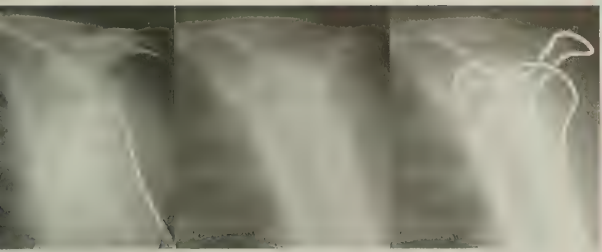
Kocher's method: • Flex the elbow and put traction on the arm.

- Apply forced external rotation and adduction of the point of the elbow across the chest while the arm is flexed at the shoulder.
- Now apply internal rotation, so that the volar aspect of the forearm lies on the chest. Risk: fractures of the humerus.

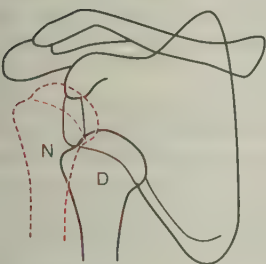
Scapular manipulation: (good, if you can palpate the lateral border of the scapula.) Drugs to relax muscles aren't needed in >60%. medline:877953 It is successful in 90%. medline:1416331 Have the patient sit with the uninjured shoulder against a wall. Ask a helper to face the patient and lift the outstretched arm on the affected side to horizontal. The assistant places the palm of his free hand against the midclavicle on the injured side, to give forward traction to the humerus, while you manipulate the scapula by adducting the inferior tip using your thumb, while stabilizing the superior aspect with your upper hand. Get post-reduction radiographs.

Surgery may be needed for recurrent dislocation (eg the Putti–Platt and Bankart reinforcement of the capsule and rotator cuff).

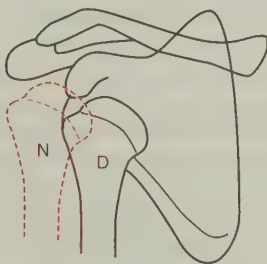
Posterior dislocation of the shoulder is rare, and may be caused by seizures. It may be hard to diagnose from an anteroposterior radiograph ('light-bulb' appearance of humeral head); lateral radiographs are essential. Refer to an orthopaedic surgeon.



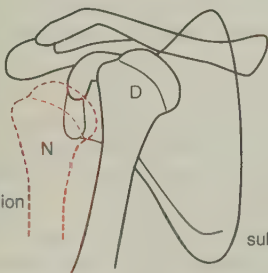
anterior dislocation of the shoulder and a post-reduction image that is copied and highlighted. The dislocation can be painful, so positioning is difficult for the radiographer. These are attempted lateral views of the capula. After reduction, the head of the humerus lies in the centre of the 'Y' with the coracoid process anterior and the acromion posterior.



subglenoid



subcoracoid
(most common)



subclavicular

N = normal position
D = dislocated

Fractures of the elbow and forearm

The humerus *Supracondylar fracture*: (eg a child who has fallen on an outstretched hand). Keeping the elbow in extension after injury prevents brachial artery damage. Median & ulnar nerve palsies are rare complications. Some may be reducible under GA, but open reduction & internal fixation (eg with K-wires) may cause fewer complications, more stable fixation, and better anatomical reduction (for type III supracondylar fractures). medline:10968183

Avoid flexing the elbow by $>90^\circ$. Careful post-op observations are needed. The radial pulse may not return for 24h post-reduction, but limb perfusion is usually enough to prevent Volkmann's contracture (p 616). Pulse oximetry \pm Doppler helps assess vascular compromise.

Fracture of the medial condyle: These may require surgery if manipulation fails to bring about satisfactory reduction.

Fracture of the lateral condyle: Surgical fixation will be required. Complications: cubitus valgus and ulnar nerve palsy.

T-shaped intercondylar humerus fracture: This is a supracondylar fracture with a break between the condyles. The presence of a fracture is suggested by the hoisting of the sail-shaped fat pad on the anterior aspect of the distal humerus (the 'fat pad sign'), seen on lateral elbow radiographs—which indicates effusion. Compare with the other elbow. If no fracture is obvious but an effusion is present, treat initially with a broad arm sling. Re-x-ray after 10 days (when fractures are more easily seen): if clear, start mobilization. For fractures, internal fixation may be needed. Physiotherapy and early mobilization are vital in preventing stiffness. medline:10588476

Fractures of the radius head The elbow is swollen and tender; pronation & supination hurt. Radiography often shows an effusion, but minor fractures are often missed. Undisplaced fractures can be treated in a collar and cuff sling—if the fracture is displaced, internal fixation or excision of the radial head may be needed. Complications: radial nerve palsy (rare).

Pulled elbow Typical patient: 1–4yr-old who has been lifted by the arms in play, causing the radial head to slip out of the annular ligament. Elbow rotation (forced supination with a thumb over the radial head) may be all that is required, producing a click on reduction. Radiographs are not required. Immobilizing the elbow for 2 days after manipulative reduction improves chances of success. N=64
medline:10853908

Elbow dislocation A fall on a not yet fully outstretched hand, with elbow flexed, causes posterior ulna displacement on the humerus, and a swollen elbow, fixed in flexion. Brachial artery and median nerve damage are rare.

Reduction (\pm GA): stand behind the patient, flex the elbow, and with your fingers around the epicondyles, push forwards on the olecranon with your thumbs, and down on the forearm. Hearing a clunk heralds success. This may be aided by traction at the wrist from an assistant standing the other side of the trolley. Immobilize in a sling for 3 weeks. *Complications*: Stiffness, instability, ectopic ossification, radioulnar joint disruption. medline:969293

Olecranon fracture There may be wide displacement of the proximal fragment, caused by triceps pull. Open reduction with internal fixation, eg tension band wiring (TBW) is often used. Post-op loss of reduction (causing significant articular step-off or gap) occurs in up to ~50%. medline:144644

Monteggia and Galeazzi fractures See p 754 & p 748.

Elbow arthroplasty In comminuted distal humerus fractures, open reduction & internal fixation may be difficult, eg in osteoporotic bones. Hence the occasional use of total arthroplasty as a primary procedure. medline:1108415

Classification of supracondylar fractures:

There are 2 big categories: *extension* (95%) and *flexion* types. Gartland has classified extension fractures thus: www.medmedia.com/orthoo/91.htm

- Type I: non-displaced fractures
- Type II: displaced with intact posterior cortex
- Type III: posterior displacement. IIIA is posteromedial and threatens the radial nerve. Type IIIB is posterolateral and threatens the median nerve (esp. the anterior interosseous branch, which innervates deep flexors to the index finger as well as flexor pollicis longus; it is an entirely motor nerve—so injury is easily missed). www.txt.co.uk/full.cfm?1084

Reduction of a fracture of the distal radius (Colles' type)

Before

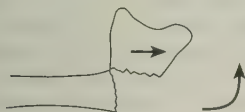
(radius shown alone in lateral view only)



Dorsally angulated
Dorsally displaced
Loss of ulna tilt
+/- Impacted

1. Traction should be applied to the hand with an assistant to provide counter-traction at the elbow. The fracture can often be felt to disimpact with 'clunk'.

2. Exaggerate the dorsal angulation while maintaining distal traction. This is most easily done by keeping both thumbs on the dorsal fracture line. The intact dorsal periosteum will prevent over-distraction.



3. Correct dorsal and radial angulation, again maintaining distal traction. Aim for anatomical alignment.



4. Apply plaster of Paris (POP), moulded to provide 3-point pressure. Traction should be maintained while the POP is applied. This is most easily done by pulling on the ring and little finger against counter traction. This applies both palmar and ulnar forces.

- An inability to achieve a good position may indicate soft tissue interpositioning.
- Finding good staff with experience saves remanipulating in theatre.

Fractures and dislocations of the wrist

Distal radial fractures—eg Colles' (common in osteoporotic postmenopausal ladies who fall on an outstretched hand). There is backward angulation and displacement producing a 'dinner-fork' wrist deformity when viewed in pronation (the fingers are the prongs). Avulsion of the ulna styloid process may also occur. If there is much displacement reduction is needed (esp. if backward and proximal shift of the distal fragment). In comminuted fractures, cast immobilization can be supplemented with pins and plaster technique and external fixators—or internal fixation. medline:972815

Treatment: Bier's block (regional anaesthesia)—method:

medline:9413772

▶▶ Never use bupivacaine for a Bier's block (cardiotoxic if the cuff is inadvertently deflated). The use of a double cuff is controversial—a properly applied and checked tourniquet should suffice.

- 1 Place a loose tourniquet around the upper arm.
- 2 Empty the arm of blood either by raising above the heart for 1 min or by the use of an esmarch bandage.
- 3 Inflate the cuff to 100mmHg above the systolic BP.
- 4 Inject 30–40mL 0.5% prilocaine into a vein on the back of the hand.
- 5 Allow anaesthetic to develop (20–30min), then manipulate the fracture.
- 6 30min after the injection deflate the tourniquet. NB: Sudden early release of prilocaine into the circulation can cause fits. Other methods such as direct infiltration of the haematoma (haematoma block) are less effective. The alternative is general anaesthesia.

The manipulation: Prepare a plaster back slab up to the knuckles. Ask an assistant to hold the elbow. Apply traction to disimpact the fragment and push it forwards, and towards the ulnar side. Keeping the arm under traction, apply the slab, with wrist slightly flexed and hand in ulnar deviation. Support in a sling, once a radiograph has shown a good position. Check radiologically in 5 days, when swelling has reduced. The plaster is then completed. Complications: median nerve symptoms (should resolve after good reduction); ruptured tendons; malunion; Sudeck's atrophy (p 695). NB: a right Colles' plaster is unlikely to affect driving. Injury 2002 33 55

Smith's fracture (Distal radius, radial fragment angled forward). Manipulate, with the forearm in full supination. Fixation is commonly needed.

Bennett's fracture Carpometacarpal fracture/dislocation of thumb. *Rx*: percutaneous wire fixation. Exact reduction ↓ risk of future OA. medline:8077832

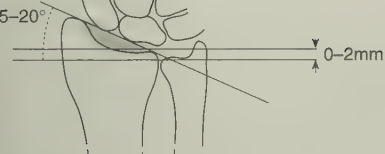
Scaphoid fracture (Common and easily missed on radiography; results from falls on the hand). **Signs:** Swelling; pain; tender 2cm distal to Lister's tubercle of the radius, and on proximal pressure on the extended thumb or index finger. **Images:** Get oblique 'scaphoid' radiography. If –ve, and fracture is likely, long-axis CT is wise (also identifies unstable fractures). If unavailable, put in plaster and x-ray at 2 weeks, when the fracture may be visible. Nondisplaced fractures involving the waist or the proximal pole are often treated with immobilization in a long-arm thumb-spica cast for several weeks, then in a short-arm thumb-spica cast until union. Percutaneous cannulated Acutrak screw fixation may be better. www.ejbs.org/cgi/content/full/83/4/483

Complications. As the nutrient artery enters distally, there may be avascular necrosis of the proximal fragment, causing late wrist degeneration.

Wrist dislocation (eg scapho-lunate or luno-triquetral) May be anterior or posterior. Manipulation and often open reduction, and plaster immobilization eg for 6 weeks. Median nerve compression may occur. medline:1087422

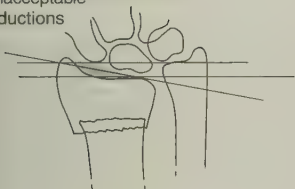
-P views

Normal



The articular surface of the radius is level with, or proud of, the ulna and is tilted towards the ulna.

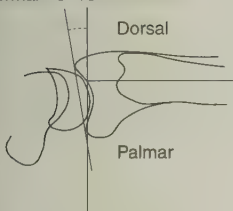
Inacceptable reductions



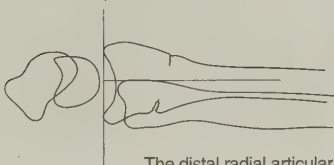
Note loss of radial length and reduction in ulnar tilt of the radius.

Lateral views

Normal 0-10°

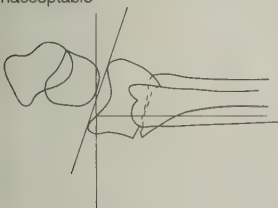


Acceptable



The distal radial articular surface is vertical

Inacceptable



The distal radius surface is dorsally angulated. If allowed to heal in this position it will cause a marked restriction in function.

Injuries to the hand

► There are no minor injuries to the hand. Any breach of the integument may be the start of a chain of events that leads to loss of our most useful appendage. Contrary to beliefs of poets, our fingers are not protected from accidents by an invisible ring of shining:* they are very frequently injured.

Infections Strep and staphs are typical causes (and mixed Gram -ve in the immunocompromised). Spread may occur within a tendon sheath which may slough. Injuries in diabetics need special vigilance.

Treatment: If pus is present, take a swab. Give flucloxacillin 250mg/6h po or erythromycin 500mg/12h po starting 1h before surgery. Under GA and tourniquet control, the bloodless field is explored. If necessary, the synovial sheath is excised at its proximal end. Local collections of pus can occur in tendon sheaths or palmar spaces. Surgical drainage ± irrigation is indicated. Any abscess (eg paronychia or subcutaneous) or infected fascial spaces must also be drained and cleaned. Sometimes primary suturing is possible, provided all necrotic material is removed. A chronic 'cold' abscess of the tendon sheath suggests tuberculosis. Enlist expert help early.

medline:988489

Nerve injury (see p 664 & p 720) Examine sensory and motor function. Inability to lift the thumb out of the plane of the palm ≈ median nerve injury (failure of abductor pollicis brevis). Nerves smaller than the digital nerve at the level of the distal interphalangeal joint are rarely repairable. Function is better if the injury is to a nerve subserving a single modality (motor or sensory), as regenerating fibres simply grow down the nearest Schwann tube (at ~1mm/day) irrespective of distal function. Let an expert decide the method of repair: primary, secondary, or nerve graft.

medline:1069657

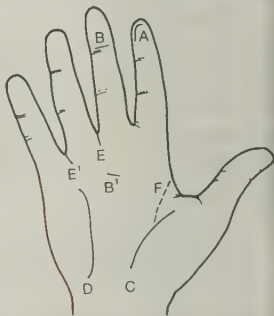
Tendon injuries Failure to raise the affected finger by extending the MCP joint ≈ extensor tendon division. 75% are closed injuries.

medline: 1079823

Failure to flex the DIP joint against resistance ≈ divided flexor digitorum profundus. If this is intact, but flexion of the PIP joint is affected, there is division of superficialis. Flexor pollicis longus section leads to inability to flex the interphalangeal joint of the thumb. In general, flexor tendon injuries are best treated by primary repair (most are open injuries). If there is loss of tendon substance, or delayed presentation, a staged repair with a silastic implant to keep the tendon sheath open, followed by a tendon graft, may be needed. Intensive hand physiotherapy with supervision is essential.

702

Hand incisions: A, pulp space drainage; B & B', lavage of flexor tendons; C, drainage of radial bursa; D, drainage of the ulnar bursa; E & E', for the mid-palmar space; F, drainage of the thenar space



Stephen Spender 1985 *To My Daughter*
Circle this bone with shining...

B... clasp of her whole hand around my finger
 My daughter, as we walk together now
 All my life I'll feel a ring invisibly
 Circle this bone with shining...

limb surgery in bloodless fields

This may be achieved with pneumatic tourniquets, which must have adequate width and curve, and be applied for as brief a time as possible. medline:7894409

Bruner's rules: These minimize risk of ischaemic limb changes.

Width of tourniquet: 10cm for the arm; 15cm for legs.

Apply to the upper arm, or mid/upper thigh.

Use ≥ 2 layers of orthopaedic wool to provide adequate padding (make sure it does not get wet with the skin preparation fluid, which should be aqueous, so that if wetting happens inadvertently, 'burns' do not occur).

Inflation—arm: 50–100mmHg above systolic BP; leg: double systolic.

Deflation—must be within 2h.

Re-application after only brief reperfusion is inadvisable.

medline:3396547

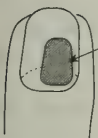
Avoid heating the limb (cooling is better, if feasible).

medline:8734236

Apply only with the utmost caution to an unhealthy limb.

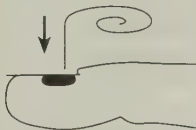
Ensure the apparatus is calibrated weekly, and is well maintained.

Document duration and pressure of tourniquet use.



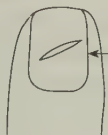
Sub-ungual haematoma

Release with needle by 'drilling' down – twiddling it between finger and thumbs; or by heating a paper clip to white heat and burning a hole through the nail.



If left, the blood does not clot for up to 24hrs, therefore there is a throbbing pain due to pressure. This sometimes works (relieving the pressure) up to 2 days after the original injury.

Beware: subungual lacerations to the nail bed



Nail removed

Remove the nail first—then stitch.

Also remove the nail if there is nail avulsion.

Fractures in the hand

Fractures of the metacarpals These only need manipulation if there is gross displacement or angulation. The 5th metacarpal is most commonly involved, often from a punching injury. Immobilization for up to 10 days in a wool & crêpe bandage may be all that is needed, with finger movement encouraged after 2–3 days. Closed reduction + cast immobilization is also used.

medline:10961550

Longer periods of splinting in a plaster or 'boxing glove' bandage can result in a stiff hand. Refer any fractures with obvious rotational deformity (a clinical, not a radiological decision), as this can be disabling. Rotational fractures disclose themselves by producing a rotation of the fingers (flex the fingers and look at all the nails end on); they usually require operative fixation with plate and screws, as do fractures of ≥ 2 metacarpals. ► Beware wounds overlying metacarpophalangeal joints—these are often from the teeth of the punched victim, are contaminated bites, and may communicate with the joint.

Fractures of the proximal phalanx Spiral or oblique fractures occurring at this site are likely to be associated with a rotation deformity—and this must be corrected. Often, the only way to do this accurately is by open reduction and fixation, eg with a single compression screw.

medline:8409671

Fractures of the middle phalanges Manipulate these; splint in flexion over a malleable metal splint, strapping the finger to its neighbour (buddy strapping). The aim is to control rotation, which interferes with later finger flexion.

Fractures of the distal phalanges may be caused by crush injuries and are often open. If closed, symptoms may be relieved by trephining the nail to reduce swellings. Split skin grafts from the thenar eminence may be needed for partial amputations of the finger tip. If subluxation is a problem, joint stiffness may be avoided by open fixation.

medline:7275291

Mallet finger The tip of the finger droops because of avulsion of the extensor tendon's attachment to the terminal phalanx. If the avulsed tendon includes a piece of bone, union is made easier—using a special splint, eg with 0° of extension. Use for 6 weeks.

medline:10746471

Interphalangeal arthrodesis may be needed if active extension remains limited. Poorer outcome is associated with delay in splinting, and if >50yrs old.

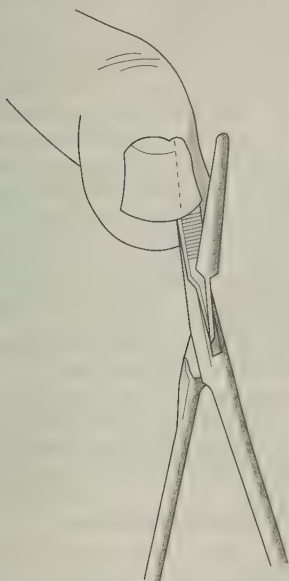
Gamekeeper's thumb This is so-called because of the rupture of the ulnar collateral ligament of the metacarpophalangeal joint of the thumb during the forced thumb abduction that occurs when wringing a pheasant's neck. The same injury is described in dry ski-slope participants who fall and catch their thumb in the matting ('Hill-end thumb'). Diagnosis can be difficult as the thumb is so painful to examine, but to miss this injury may condemn the patient to a weak pincer grip—inject 1–2mL 1% plain lidocaine (=lignocaine) around the ligament to facilitate examination. Differentiation of complete vs partial tears of the ligament is crucial because the treatment for complete tears is surgical. Radiographic evaluation, while important, has a limited rôle. Small-joint arthroscopy is a new technique which may have a rôle in planning repair of full tears. Very often the ligament ends are turned into the joint.

medline:8623701

Partial tears, or those associated with uncomplicated avulsion fractures of the proximal phalanx, can be adequately treated using simple shortarm thumb spica casting.

Nail avulsion

1. Warn the patient the nail may not regrow or regrow properly.
2. Ring block the finger and test it is numb.
3. Push one jaw of a small straight clamp down one side of the nail at its edge.
4. Close the clamp firmly. Warn the patient to look away.
5. Turn the clamp on its long axis as if opening a sardine can. The nail will wrap round the clamp and peel off its nail bed.
6. Having repaired the nailbed, dress the wound with some paraffin gauze, tucking some into the pocket at the base of the nailbed.



Pelvic fractures

Rang's aphorism: *The pelvis is like a suit of armour: after damage there is much more concern about its contents than about the structure itself.* Owing to the ring structure of the pelvis, single fractures are often stable and just need a few weeks rest. In contrast, ≥ 2 fractures in the pelvis (with one above the level of the hip) renders the ring unstable and is a serious injury, with internal injuries in 25%. The force producing the fracture is usually compressive, eg from a traffic accident (60%) or fall. Signs to look for include bruising, perineal or scrotal haematoma, and blood at the urethral meatus.

Examples: Fractures of the ilium and pubic ramus (on one or both sides); fracture through a sacroiliac joint and a pubic ramus, as below.

Malgaigne's fracture (20% of all pelvic fractures, 60% of unstable ones): disruption of the pelvis anteriorly and posteriorly with displacement of a fragment containing the hip joint.

Acetabular fractures Common sites: posterior lip or transverse. Two 45° oblique radiographs (\pm CT scans) are needed to define injuries exactly (easy to miss on x-ray). Treatment: open reduction and reconstruction of the articular surface—to delay the onset of secondary osteoarthritis.

Examining patients with suspected pelvic fracture Gentle palpation of the iliac crests and the pubis, or gentle inward compression of the iliac wings is often enough to elicit pain in an awake patient. Avoid the outmoded pelvic rock test (iliac compression while pressing on the symphysis pubis; it worsens hemorrhage, disrupts nerves, and may cause soft tissue damage. Once a pelvic fracture is suspected, avoid manipulations. The aim is to minimize pelvic motion. www.merginet.com/tgp/2000/0009/feature-pelvic.shtml

Complications • Haemorrhage (OPPOSITE, eg internal iliac artery). Check foot pulses, BP, CVP + urine output often. Transfusion is often needed.

- Bladder rupture—may be intraperitoneal or extraperitoneal.
- Urethral rupture, often at the junction of the prostatic and membranous parts in males. The appearance of a drop of blood at the end of the urethra is suggestive. He may be unable to pass urine (avoid repeated tries). On rectal exam, the prostate may be elevated out of reach. NB: CT is the image of choice in trauma patients with haematuria.
- Vaginal and rectal perforation—look for bleeding. Both are rare.
- Trapping of the sciatic nerve—there is persistent pain.

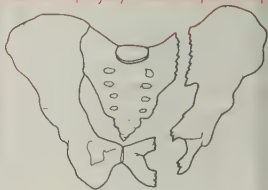
Treatment Relieve pain and replace blood (OPPOSITE). If urethral rupture is suspected, check with a urethrogram before catheterizing; a suprapubic catheter may be needed. Get urological help. A small volume of urine suggests bladder rupture. A cystogram or CT is needed. If no pelvic fluid is seen on CT, bladder rupture is unlikely.

medline:11487073 &: 1080361

Avoid urethral catheters: they may make a false passage.

Reassuring signs on a pelvis radiograph. This sort of injury needs expert help

- Symphysis pubis separation <1 cm
- Integrity of superior & inferior rami
- Integrity of acetabula & femoral necks
- Symmetry of ilium & sacroiliac joints
- Symmetry of the sacral foramina, eg by evaluating the arcuate lines
- No fracture of transverse process of L5



Shock and pelvic fracture

• This carries significant mortality (14–55%; worse if base excess on arterial blood gas analysis is ≤ -5). medline:10966029 & 10963504

• *Is the patient pregnant?* This poses big problems, eg ↑pelvic blood flow. Resuscitate vigorously and meticulously: p 684.

• Ways to reduce blood loss: avoid manipulations of the pelvis. Internally rotate both lower legs to close an 'open-book' fracture. Apply a pelvic binder. Suspend the patient in a pelvic sling promptly. The patient lies supine with the pelvis over the sling's webbing, which exerts upward and medial (thereby compressing) traction via weights and runners suspended above the bed. Traction is also applied to the legs. Surgical reconstruction may then be undertaken.

• Alternative: immediate Hoffmann's external fixation frame. medline:10392195

• Look for associated abdominal and pelvic injury. In one series of pelvic fractures there were splenic lesions (37%), diaphragm (21%) and bladder ruptures (24%), liver lacerations (19%), urethra lesions (17%), intestinal lesions (17%), and ruptures of the kidney (9%). medline:10025036

• Sometimes diagnosis is hard. Prompt spiral CT identifies those needing a special procedure, such as angiographic embolization. medline:11015102

• Ordering of interventions matters—eg laparotomy if indicated, intra-operative fixation, followed by postoperative embolization. medline:9731815

• Inter-disciplinary cooperation is vital—and must be practised.

Injury to the hip and femur

Incomplete or impacted femoral head fractures may present with coxa valgus; complete fracture with dislocation may present with coxa varus.

Intracapsular fractures occur just below the femoral head. This often causes external rotation and leg adduction. The injuring force may be trivial and the patient may be able to walk (but with difficulty). As the medial femoral circumflex artery supplies the head via the neck, ischaemic necrosis of the head may occur, particularly if there is much displacement. Femur fractures fill 20% of all UK orthopaedic beds. The incidence is rising (~1:100/yr in females aged 75–84). Mortality: 50%.

Treatment: Assess vital signs. Treat shock with Haemaccel®, but beware incipient heart failure. If present, monitor CVP.

- Relieve pain (eg morphine 0.2mg/kg IM + prochlorperazine 12.5mg IM).
- Images: a good quality lateral is essential to make the diagnosis if there is impaction or little displacement. 5% can be missed unless CT is used.
- Prepare for theatre: FBC, U&E, CXR, ECG, crossmatch 2U, consent. Tell the anaesthetist about any drugs the patient is taking.
- Sort out any medical problems before embarking on surgery.
- If displacement is minimal, use either multiple screw fixation or Garden screws. The head is excised and a prosthesis inserted if risk of failure of internal fixation is high (eg age of patient; state of bones). medline:1104157

Intertrochanteric–extracapsular fractures (between greater and lesser trochanters). They occur in a younger age group and, as blood supply is adequate, non-union is rarer. **Treatment:** Compression screw plate. The principle of the screw plate is to fix the fracture, but to allow compression by sliding. Surgery is associated with ↓length of hospital stay and improved rehab. Conservative treatment may be acceptable if modern surgical facilities are unavailable, and will result in a reduction in complications associated with surgery, but rehabilitation is likely to be slower and limb deformity more common. medline:11034683 &: 116869

Femoral shaft fracture ▶ Is the femoral artery torn? (Look for swelling and check distal pulses.) Sciatic nerve injury may also occur. The proximal bone fragment is flexed by iliopsoas, abducted by gluteus medius and laterally rotated by gluteus maximus. The lower fragment is pulled up by the hamstrings and adducted (with lateral rotation) by the adductor. **Treatment:** Typically, this is with a locked intramedullary nail introduced proximally over a guide wire that is manipulated across the fracture under fluoroscopic control. This allows early mobilization. Alternatively, manipulation under anaesthesia (exact reduction is not needed) with fixed traction with a Thomas knee splint, or skeletal traction, or sliding traction, with the thigh supported on a frame, or a cast brace with a hinge at the knee (this permits early weight bearing). Union takes 3–6 months.

Condylar fractures & tibial plateau fractures Being intra-articular, these demand accurate joint reconstruction to minimize later OA (p 668).

Posterior hip dislocation (eg to front-seat passengers if the knee strikes the dashboard). Feel for the femoral head in the buttock. The leg is flexed, adducted, and short. The sciatic nerve may be lacerated, stretched, or compressed; early MRI diagnosis may prevent later equinus foot deformity. **Treatment:** Reduction under GA, by lifting the femoral head back into the joint. Traction for 3 weeks promotes joint capsule healing. medline:109431

Fracture dislocation of the hip This is described on p 706.

Preventing hip fractures

Prevent falls: eg good lighting, less sedation, & keep-fit programmes.
Teach exercise and balance training (physiotherapy or Tai Chi).
Prevent osteoporosis: eg exercise, HRT, bisphosphonates, p 676.
Ensure good vit D intake (plasma levels $\geq 30\text{nmol/L}$; esp. in northern climes). Vitamin D lack is associated with hip fracture in an important way, whether or not patients are osteoporotic. www.txt.co.uk/full.cfm?1085
Meta-analyses suggest that hip protectors decrease risk of hip fracture in selected populations at high risk. Acceptability by users remains a problem, due to discomfort and practicality. meta-analysis medline:11034706

The following may prevent complications after hip injury

Early mobilization to prevent thromboembolism. meta-analysis
Co-ordinated multidisciplinary inpatient rehabilitation. medline:11103467
Good nutrition—but meta-analyses do not provide much support for specific multi-nutritional commercial food supplements. meta-analysis medline:1103731

Injuries of the lower leg

The patella Lateral dislocation may result from a blow to the side of the knee, causing a tear in the medial capsule and the quadriceps aponeurosis typically in the young. Knee flexion is limited. Recurrent dislocation may be related to under-development of the lateral femoral condyle.

Sudden contraction of quadriceps may snap the patella transversely whereas direct force may produce a multiple 'star-shaped' fracture (undisplaced within the quadriceps tendon).

The knee *Injury to a collateral ligament* is common in sport, eg presenting with effusion \pm tenderness over the ligament injured. Rest is needed then firm support. If there is complete rupture, varus and valgus strain opens the joint if the knee is held in 10° flexion (and if the knee is held in extension, if a cruciate ligament is also torn). Surgery to isolated medial collateral ligaments is rarely warranted. medline:111044

Anterior cruciate tears: Typically, these follow posterior blows to the tibia or rotation injuries, when the foot is fixed to the ground. Signs: effusion, haemarthrosis, and a +ve 'draw' sign. (Immobilize the patient's leg by sitting on his foot, and then with the knee in 90° flexion, grip the upper tibia, and try to draw it towards you, away from the femur. Note: the reverse 'sag' back' suggests a posterior cruciate tear—eg after car accidents as the knee strikes the dashboard.) Do a 'pivot shift test' (p 634). Examination under stress may be needed. **Treatment:** This is problematic. 3 weeks' rest in a plaster cylinder + physio may help. If knee instability and giving way persist, consider autografts, allografts, or synthetic graft replacements. medline:108824

Semilunar cartilage (meniscal) tears: Medial cartilage tears (eg 'bucket-handle') follow twists to a flexed knee (eg in football). Adduction + internal rotation causes lateral cartilage tears. Extension is limited (knee locking) as the displaced segment lodges between femoral and tibial condyles. The patient must stop what he is doing, and can only walk on tiptoe, if at all. The joint line is tender, and McMurray's test is +ve (p 634). If the 'handle' of the 'bucket' becomes free at one end ('parrot beak' tear), the knee suddenly gives way, rather than locking. MRI gives tear location, morphology, length, depth, and stability, and helps predict tears requiring repair. medline:108199

Tibia fracture *Avulsion fractures* of the intercondylar region often occur with anterior cruciate injury. Arthroscopic reduction may be tried. medline:75526 Sliding traction, or surgery with open elevation of the joint surface may be needed. Open fractures of the shaft are common as there is so little anterior covering tissue. Tearing of the nutrient artery may make non-union a problem in lower tibia fractures.

With closed fractures, intramedullary nailing may be sufficient.

Open fractures: One option is one-stage debridement + stabilization by locked intramedullary nailing \pm bone grafting from iliac crest \pm making good of skin loss using free muscle flaps, eg from latissimus dorsi or gracilis. medline:109294

Isolated fibular fracture A supportive dressing may be sufficient.

Pretibial lacerations The shin (particularly if elderly) has a poor blood supply. It is vulnerable to flap wounds, eg caused by steps of buses. **Treatment:** Try hard to iron out all the flap, and reposition it carefully: a good ideal tool is the 'wrong' end of a Vacutainer® needle, sheathed in its rubber case. The important thing is to prevent tension (tension \rightarrow breakdown \rightarrow plastic surgery)—so skin closure with adhesive strips (Steristrip®), which can be loosened if the tissues swell, is better than sutures. Dress the wound and advise a support bandage, and leg elevation. Review to check for infection, wound tension, and necrosis.

Gustilo classification of open fractures*

Type I Low-energy wounds <1cm long, eg caused by bone piercing skin.

Type II The wound is >1cm—typically from a higher energy wound, causing moderate soft tissue damage.

Type III Extensive soft tissue injury, which is usually >1cm long. Fractures from high speed gunshot injuries are always Type III, as are most crush injuries from fast vehicles. Fractures with significant diaphyseal bone loss are generally Type III. Type III fractures are denoted IIIB if there is extensive stripping of bone, with inadequate coverage. Type IIIC implies arterial injury needing repair. (IIIA implies that there is enough soft tissue to provide adequate local coverage.)

*Scott Kozin *Handbook of Common Orthopaedic Fractures*, Medical Surveillance Inc



Before
Flap retracts back



Good
No tension in flap.
Held with adhesive
sutures.



Bad
Flap stretched too
tight, and likely to
necrose.

Ankle and foot strains, and fractures

Ankle ligament strains Twisting inversion accounts for the common strain to the anterior talofibular part of the lateral ligament. **Signs:** Stiffness, tenderness over the lateral ligament, pain on inversion. Only if there is swelling or inability to walk (eg 4 steps) do radiographs to rule out lateral malleolar fracture (p 734). **Treatment:** See OPPOSITE.

Medial deltoid ligament strains are rare, and often need simple strapping only. Supervised walking exercises and diadynamic therapy relieve pain. NB: minor sprains may respond to rest and simple analgesia ± topical NSAIDs (ibuprofen, ketoprofen, piroxicam). **Prevention:** Footwear newness is said to play a more important rôle than shoe-heel height. Proprioceptive training can ↓ incidence of ankle sprains in athletes. medline:11086757

Malleolar and metatarsal fractures Either malleolus may be fractured by the above injuries. Rotation causes oblique lateral malleolar fracture (hard to see except on lateral radiographs), or a proximal fracture of the 5th metatarsal (avulsion by peroneus brevis). **Treatment:** Surgery to restore the length of the fibula by open reduction and fixation of the malleoli.

Eversion injuries may produce bimalleolar fractures. These are very unstable and will either require an above-knee plaster (with the knee in flexion to control rotation) or internal fixation. image -
www.txt.co.uk/full.cfm?1086

Dupuytren's fracture: Distal fibula fracture + distal rupture of tibiofibular ligament ± fracture of the back of the lower end of the tibia (trimalleolar) with lateral displacement of the talus. **Treatment:** Open reduction + screw fixation if > 1/3 of the joint surface is involved; no full weight-bearing for 7 wks.

Maisonneuve's fracture: Proximal fibula fracture + syndesmosis rupture, and medial malleolus fracture or deltoid ligament rupture. If 2 bones dislocate where no true joint exists, the term diastasis is used. ▶ Always examine the proximal fibula with 'ankle sprains'. Treatment of Maisonneuve's fracture is operative, with restoration of fibular length, repair of ligaments, and placement of one or two suprasyndesmotomic screws. medline:10917243

Lisfranc fracture-dislocation at the 1st tarsometatarsal joint: A commonly missed fracture in multitrauma patients, it can also occur by stepping awkwardly off a kerb. It may cause compartment syndrome of the medial foot (± later arthritis and persistent pain). medline:11035858 **Images:** Plain x-rays: look for widening of the gap between the medial cuneiform and the base of the 2nd metatarsal. Because of the overlapping bones, subluxations can be hard to see. CT helps. medline:10584818 **Rx:** Achieve precise anatomic reduction. Open reduction and temporary screw or K-wire fixation may be best. medline:9265797

Other fractures **Fractured neck of talus** can occur after forced dorsiflexion, and is a serious injury because interruption of vessels may lead to avascular necrosis of the body of the talus. **Treatment:** Closed or open reduction.

Os calcis fractures (often bilateral, following serious falls; look for associated spinal fracture). **Signs:** swelling; bruising; inability to weight bear. ▶ Does the fracture enter the subtalar joint? Only opt for conservative treatment in extra-articular fractures, or minor displaced intra-articular fractures in nonambulatory patients. Otherwise, get expert surgery. Anatomic reduction is achievable in >50% of even 3-part fractures. medline:1023203

Mid-tarsal (eg calcaneocuboid) dislocation produces a painful, swollen foot. The navicular proximal articular surface may not articulate with the talus. **Treatment:** Early manipulation + plaster cast for, eg 7 weeks. medline:272106

2nd metatarsal march fracture follows stress, eg from marching. **Signs:** inconstant. **Imaging:** bone scan hot spot. **Rx:** plaster cast for 6 weeks. medline:903554

Fractures of the toes: Simple protective dressings usually suffice.

Treating ankle inversion trauma when there is no fracture

Compression bandage, or below-knee plaster if there is much swelling. Rest with leg elevation.

Ankle movements (especially dorsal extension).

medline:10560560

Weight-bearing as soon as the pain allows. A few days later, when pain and swelling subside, examine to detect lateral ligament rupture.

If you suspect ligament rupture, consider stress radiographs (GA may be needed). Look for talar tilt \pm anterior talar displacement, in comparison to the unaffected side. medline:10431302 Be sceptical of the result: meta-analyses paint a pessimistic picture of reliability.

medline:10336051

If cost allows, MRI is a non-invasive and accurate way to evaluate injured lateral collateral ligaments.

medline:9891772

Treat simple lateral ankle ligament tears non-operatively. Use surgery for dislocated bone avulsions and chondral or osteochondral fracture.

Conservative treatment of ligament rupture: immobilization of fibular ligaments with as little compromise of ankle joint function as possible, and rehabilitation with muscle strengthening and proprioception training.

Recurrent tears in athletes will probably need surgery.

At surgery, ligament stumps are reapproximated in anatomic position, reinforced with local tissue if needed.

medline:10431302

Classification of malleolar fractures:

There is the **Weber** and the **AO** system. The **AO** system is detailed in the figure.*

1 Transverse fibula fracture (#) at or below joint line.

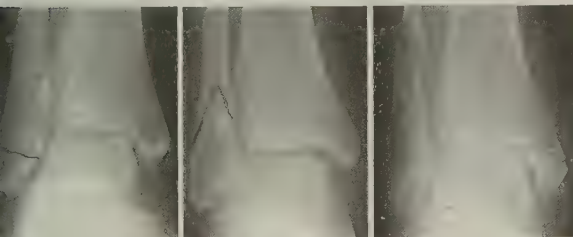
2 Spiral fibula # starting at the joint line \pm medial injury.

3 Oblique fibula # above a ruptured tibiofibular ligament + medial injury.

4 Maisonneuve's fracture, only the fibular fracture is more proximal.

Both systems have problems.

medline:10209586 & 10897286



►► Head injury

Emergency care As always, treat the **ABCs**—Airway, Breathing, and Circulation. ►Note pupil sizes and reactivity. ►Give O₂ and treat shock. ►Protect the cervical spine. ►Do not neglect other injuries. Primary brain damage occurs with the initial trauma. The clinical imperative is to prevent or ameliorate secondary brain injury, ie further neuronal death.

Ask about: Time and mechanism of injury • Trends in consciousness level

- Symptoms of co-existent medical conditions (especially epilepsy, diabetes). Any alcohol or drugs? **On examination:** Look for:
 - Palpate **scalp & skull** for evidence of compound or depressed fracture.
 - Examine the **facial bones** including orbits and middle third.
 - Examine **cervical spine** for tenderness, deformity, and abnormal neurology.
 - Check **ears & nose** for bleeding or CSF leaks. The presence of these or blood behind the tympanic membrane may indicate a fractured base of skull. Bruising over the mastoid (Battle's sign) and periorbital bruising ('Raccoon eyes') are late and unreliable signs of fractured base of skull.

Neurological assessment Write full notes; record times.

- Assess the **Glasgow Coma Score** (GCS, p 680) accurately and repeatedly. Record all 3 parameters (**Motor**, **Eyes**, and **Verbal**). If there is a difference between the sides, record the better result. If there is the possibility of a spinal injury, check for a response such as grimacing to a painful stimulus applied above the clavicles. Assess and record any contribution to mental status from alcohol or other drugs.
- **Pupils**—check size and reactivity every few mins until stable. Unequal pupils are less important if conscious, but a grave sign in coma.
- **Spinal cord**—look for localizing neurological long tract signs ie: assess power, tone, reflexes, and all sensory modalities, comparing right side vs left, upper body vs lower. Check for priapism and anal tone.
- **Low BP** together with an inappropriately **low pulse** rate indicates sympathetic disruption in cervical spinal cord injury.

CT scan (The best way of investigating intracranial pathology). **Indications:**

- Major head injury or minor injury with additional warning features (p 734)
- Clinical picture obscured by alcohol or drugs.
- Prior to theatre in multiply injured patients who require GA.
- Timing of CT depends on clinical circumstances. Unless decisions are immediately dependent on CT, it may be deferred to business hours.
- If immediate lifesaving surgery is needed, CT is delayed until after theatre (Skull x-rays should not be routine, particularly if an indication for CT scan exists; they don't enrich management plans or the decision to use CT; they may have a role in: suspected compound fracture head injury; possible depressed fracture or intracranial foreign body; suspected child abuse).

Cervical x-rays are needed if there has been loss of consciousness.

Pain relief Analgesia is not contraindicated in head injury; it is unethical to leave pain untreated. iv narcotic analgesia (p 678) should be titrated as required to obtain pain relief. It may be reversed with naloxone.

Further care depends on classification. **Trivial head injury:** (GCS = 15, no earlier ↓consciousness, and no post-traumatic amnesia.) **Treatment plan:**

- These patients may be discharged home after careful clinical examination. If there is any abnormality such as headache, dizziness, or vomiting, hold for observation and reassess after 6h.
- CT is not needed unless clinical problems are persistent or deteriorating.
- Note that vomiting is common after trivial head injuries in children; however, vomiting in adults is potentially more serious.

Minor head injury: (GCS = 13–15 + witnessed episode of loss of consciousness or there is significant post-traumatic amnesia or confusion.)
Management plan for minor head injury:

Treatment depends on any associated complicating factors. Always remember that 'minor' head injuries have the potential to become major head injuries. Repeated meticulous neurological examination is essential. There is a significant rate of CT abnormalities in this group and CT scan is indicated in *all* patients in whom there has been loss of consciousness or significant post-traumatic amnesia. In general, the principle to be followed in minor head injury is *CT scanning followed by selective admission*.

Admit to hospital if there is any CT abnormality, increasing headache, vomiting, facial fracture, or any other complicating factor.

Patients may be discharged home if CNS examination and CT are normal. Persistent CNS abnormalities, including confusion, or amnesia, warrant further observation for at least 6h—after which the patient must be admitted to hospital if the symptoms persist.

► **Major head injury:** Definition: GCS <13. Management plan:

If GCS ≤ 8, intubate and ventilate (following rapid sequence induction of anaesthesia, p 770). In addition, intubation may be needed for indications independent of the head injury. The aim is to maintain good oxygenation and normocarbida. Don't hyperventilate unless signs of ↑ICP (see below). Organize CT scan urgently and inform neurosurgeons.

If GCS >8, reassess GCS at frequent intervals and observe for the signs of rising intracranial pressure and brainstem herniation as follows:

- Deteriorating level of consciousness; serial GCS.
- Unequal pupils (larger pupil is ipsilateral to an expanding haematoma.)
- Abnormal respirations: recognition that the breathing pattern is abnormal is important. The exact pattern does not matter.
- Cushing's reflex is a rising BP and a falling pulse rate. This is a *late* sign and indicates imminent death. NB: bradycardia is also a feature of spinal injury but is then associated with ↓BP.

Papilloedema: another late sign and unreliable.

Rising intracranial pressure is a neurosurgical emergency and must be recognized and managed urgently.

- Perform rapid sequence induction of GA (p 770). Intubate and moderately hyperventilate aiming for a P_aCO_2 of (4–4.3kPa) (30–32mmHg). Do NOT intubate without GA—even in comatose patients, laryngoscopy and intubation without adequate sedation may raise intracranial pressure significantly for several hours. This may make the difference between a good and poor outcome. It is also essential to avoid ↓BP.

- Mannitol 20%, 1g/kg (5mL/kg) iv; repeat if CT shows subdural.

Urgent CT. NB: despite the urgency of this condition, always obtain a CT prior to theatre if at all possible. Blind burr holes may miss significant haematomas and are only indicated if there is a delay in obtaining CT.

Transfer all major head injury patients to a neurosurgical unit.

Other issues **Discharge** must be under the supervision of a responsible adult who must have written instructions. The carer must have immediate access to phone and transport. If not, admit the patient to hospital.

Combative patients: Causes: head injury, P_aO_2 ↓, P_aCO_2 ↑; alcohol. Sedation preferable to forcible restraint but will obscure the signs of head injury. Be prepared to anaesthetize and intubate (but then obtain CT scan promptly). NB: if plasma alcohol is <44mmol/L, it is an unlikely cause of coma.

►► Injuries to the face and neck

► If there is airway obstruction, summon expert help promptly. Remove blood, loose (or false) teeth, and vomit from the mouth. Lie in the semi-prone position to prevent obstruction from a swollen injured tongue, but always beware of cervical spine trauma with injuries above the clavicle. If the pharynx is swelling or there is fracture of the larynx, try gentle intubation. If impossible, do cricothyrotomy (OHCM p 754), then tracheostomy. Blood aspiration is prevented by direct pressure to the bleeding site.

Lacerations of the face Clean meticulously. Alignment of the tissues must be exact to produce a good cosmetic result.

- Complex lacerations: Is there a plastic surgeon available?
- Dog bites: give antibiotic cover (eg co-amoxiclav 1 tab/8h po).
- Rugby player's ear: aspirate haematoma (repeat every few days) and then strap orthopaedic felt pressure pads against the head. Ruptured ear drum: advise against letting water into the meatus.
- Avulsed teeth may be replaced (p 738). If inhaled, do expiratory CXR. Bleeding socket: bite on epinephrine (=adrenaline)-soaked pads, or use sutures.
- Eye injury, nose fractures, and nose bleeds: see p 514, p 550, & p 552.

Mandible injury *Signs:* Local tenderness and swelling; jaw malocclusion; a mobile fragment; bone may protrude into the mouth in open fractures; if comminuted, the tongue may cause airway problems.

Diagnosis: Orthopantomogram (OPG) radiographs. Enlist expert dental help.

Treatment of dislocations: Place your (gloved) thumbs over the back teeth and press downwards, while at the same time levering the chin upwards with your fingers (both hands). Consider midazolam sedation: titrate up to 0.07mg/kg iv: start with 2mg over 30sec, max 1.5mg if elderly—antidote flumazenil 0.2mg iv over 15sec, then 0.1mg every 60sec, max 1mg total.

Blows to the chin may cause fracture at the impact site, or indirect fractures near the temporomandibular joint.

Fractures: Open reduction + internal fixation with miniplates is better than wiring teeth together for 6 weeks. Try to arrange this for a daytime consultant list, rather than at night. Complicated fractures may benefit from lag* screw osteosynthesis.

Complications: infection; non-union.

www.txt.co.uk/full.cfm?1087

n = 492
medline:10626251

'Whiplash' injury is cervical strain caused by sudden neck extension with rebound flexion. It is common, often in rear-end collision car accidents. Hyperextension causes the damage—to the anterior muscoligamentous structures. Subsequent protective muscle spasm causes pain and stiffness, which may be severe. *Treatment:* Prompt active mobilization (if tolerated), eg with gentle stretching and posture control. Aim to prevent chronicity and 'disuse syndrome'. If symptoms last for >1yr, they are likely to be permanent. *medline:10888946* ► Do not rely on plain radiographs to rule out facet joint fractures. They are more likely if the neck is rotated at the time of forced extension. Here it may be wise to ask a senior colleague if CT is indicated.

Other car accident neck sprains (Seen in 1/3 of car users soon after accidents.) Symptoms may be delayed—and persist for years. The best treatment may be to give NSAIDs, a cervical collar, and review in the next clinic, with referral for immediate physiotherapy if symptoms warrant this. Explain to the patient that head restraints are helpful—the usual mistake is that they are adjusted too low.

*Lag compression anneals the fracture line under compressive force, giving rigid fixation and coupling of the fractured fragments to promote fusion.

Bony injuries to the face

The face forms a shock absorber which protects the brain from injury. The most common fractures to the facial bones lie along 2 hoops, from ear to ear. One is formed by the zygomatic arch, body, infraorbital rim and nose. The other is formed by the mandible.

Major blunt trauma can cause a fracture to the entire middle third of the face, which has been classified by Le Fort,^{1,2} but since the advent of seat belts these are less common.

Zygoma fractures

The Arch: Before swelling arrives, there is a depression in front of the ear and lateral excursions or opening of the mandible may be limited and painful. A suitable radiograph is the submentovertex view (SMV).

The complex: The body of the zygoma has 4 extensions: (i) Fronto-zygomatic, (ii) Arch, (iii) Maxillary buttress in the mouth, (iv) Infraorbital rim. Fractures may be palpated at these points, or disproportionately severe pain elicited on palpation. A suitable radiograph is an occipito-mental view (OM). Lateral views have no place.

Orbital floor injuries Blunt trauma around the eye can cause fracture to the orbital floor. Imaging: CT is best, but OM views may show trap door sign in the maxillary sinus. **Clinical exam for periorbital trauma:**

- Check zygomatic arch by assessing range of mandibular lateral excursions and opening. Also palpate over the arch which lies just under the skin and compare with the unaffected side.
- Patient sitting, doctor standing above and behind, place index fingers on the cheeks and look down from above for asymmetry.
- Check orbital floor:
 - diplopia on upward gaze suggests entrapment of orbital contents.
 - enophthalmos.
 - numbness in the distribution of the infraorbital nerve suggests fracture.
- Small risk of retrobulbar haemorrhage—catastrophic if missed:
 - severe pain at back of eye
 - proptosis
 - loss of visual acuity
 - prompt exam is essential.

NB: If the eye is very swollen, application of a rubber glove filled with ice is invaluable, especially if you ask a specialist to travel to check patient, they can no more open a swollen eye than you. Treatment:

Unless vision ↓, or there is significant risk of infection, admission on presentation is not mandatory: an appointment at the next fracture clinic will be adequate. If in doubt, consult with the relevant specialist. Explain about not blowing the nose in fractures in continuity with the maxillary sinus (risks periorbital emphysema and infection).

Medicolegal: Facial injuries commonly result from assault. Your notes may be used in criminal injuries claims, or as evidence in court. Often the individual is drunk and abusive, and it is late, and you are busy, but you must take a few minutes to make accurate notes (use a ruler) with diagrams. Other people will definitely take time to study and criticize what you have written. Don't forget photographs if assault is particularly serious or children involved. Document that the patient has given, or refused, permission for statements to be made to the police or legal professionals (the medical notes are confidential).

►► Abdominal injury^{1,2}

Penetrating injuries mostly require prompt laparotomy/laparoscopy.² Assess degree of penetration (if uncertain) under local anaesthetic, by extending the wound, if necessary. Ask for expert assistance with this. Laparotomy is indicated if the posterior rectus sheath is breached.

Blunt trauma Deceleration forces may tear bowel from mesentery, liver from vena cava, or bladder from bladder neck. After blunt trauma abdomen and chest may be sites of hidden blood loss—always consider intra-abdominal bleeding or pelvic fracture (p 706) if BP↓ and no source of loss is found.

Key questions: Are vital signs stable? and Is an indication for laparotomy present? (BP↓; GI, GU, or PR bleeding; evisceration; +ve ultrasound or peritoneal lavage). Liver, spleen, and kidneys are chiefly at risk—but remember the pancreas (amylase↑; injury may be missed by CT/ultrasound; endoscopy may be needed).

Tests CVP measurements, CT, and ultrasound may all mislead. There is no substitute for monitoring vital signs and examining the abdomen often. In the young and fit, signs of shock may take time to appear. Systolic BP doesn't fall consistently until ≥30% of blood volume is lost. **Radiographic implications:**¹

- Lower rib fracture (?liver/spleen trauma)
- Pelvic fracture (?rectal injury)
- Spine fracture (?renal or pancreas trauma)
- Free gas (rupture)
- Bowel displaced (?haemoperitoneum)

Signs of organ damage **Ruptured spleen:** Suggested by shock, abdominal tenderness, and distension, left shoulder-tip pain, overlying rib fracture.

Any visceral injury may cause bruising in the flanks, absent bowel sounds, and muscular spasm. **GU injury** is suggested by haematuria.

Management Maintain airway; ensure adequate ventilation. Give high-flow O₂ (12–15L/min). Treat shock (p 684). Crossmatch blood. Then:

- 1 If the patient does not respond quickly, take straight to theatre for exploratory laparotomy (after urgent CXR to rule out pneumothorax and after passing of a nasogastric tube).
- 2 Baseline tests: pulse, BP, respirations, temperature, urine output.
- 3 Look for signs of GU injury:
 - Blood at the urethral tip, signifying ruptured urethra.
 - Test the urine. Is there frank or microscopic haematuria?
 - Do a rectal exam to assess bowel integrity, presence of blood, or high-riding prostate (this suggests bladder neck injury).If any suspicious signs, seek urological help to plan urethral repair. Rarely, GU trauma needs ureteroureterostomy, ureteroneocystostomy, or even kidney autotransplantation.
- 4 Consider peritoneal lavage (OPPOSITE, p 719) or LIF diagnostic tap.
- 5 Arrange a prompt exploratory laparotomy if there is:
 - Shock (eg ruptured spleen)
 - Penetration to unknown extent
 - Peritonism (ruptured viscus)
 - Pneumoperitoneum

A non-functioning kidney on IVP is a *relative* indication. Don't deny analgesia on finishing your assessment, simply for fear of masking signs.

- 6 **Blood tests.** FBC (raised WCC may occur in ruptured spleen), U&E, amylase.
- 7 **Radiology** All with serious blunt trauma (eg RTA) need radiographs of cervical spine, chest, and pelvis as part of their primary assessment. Abdominal images are part of the secondary assessment: consider these along with IVP, thoracolumbar spine, and other injured sites.

Diagnostic peritoneal lavage (DPL)

Indications DPL is used to assist in the diagnosis of significant intra-abdominal injury following trauma, particularly when abdominal examination by palpation is unreliable (spinal injury; neuromuscular blocking drugs; head injury; alcohol) or difficult to interpret (pelvic, lower rib and lumbar spine fractures may produce abdominal pain and muscle guarding). It may be used to determine the cause of unexplained hypotension following trauma. It should be performed by the surgeon who is responsible for any subsequent operation.

Focused abdominal ultrasound for trauma (FAST) or DPL? FAST-trained surgeons achieve accuracy of >90% in detecting intraperitoneal fluid from the outset. medline:10767790 But liver & spleen tears and free fluid can still be missed. medline:10672392 Therefore, don't rely on u/s; CT may be required. n = 938
medline:11003330

CT may be difficult to interpret if there has not been enough time to opacify the bowel with oral contrast. It is good for lacerations of the spleen, liver, and kidneys. In a stable patient, CT may allow close observation of the patient, instead of, for example, splenectomy. In children CT & u/s is preferred to DPL.

Contraindications The need for urgent laparotomy is an absolute contraindication—don't delay for DPL. Relative contraindications: late pregnancy, cirrhosis, known coagulation disorder.

Technique

- The bladder and stomach must be empty, so pass a urinary catheter (not if urethral trauma) and a nasogastric tube.
- Use strict asepsis and clean the abdomen in the midline below the umbilicus. Inject lidocaine (=lignocaine) with epinephrine (=adrenaline) $\frac{1}{3}$ of the way between the umbilicus and the symphysis, in the midline (epinephrine limits false positives from abdominal wall bleeding).
- Incise down to peritoneum, then grasp the peritoneum with 2 artery forceps and lift clear of the bowel.
- Incise the peritoneum and pass a dialysis catheter into the pelvis.
- Aspirate for blood—if >5mL, DPL is positive. If <5mL, run in 10mL/kg of warm normal saline. Leave for 5–10min, then allow fluid to drain by hanging the bag below the level of the abdomen.

Interpretation Lavage fluid must be sent to the laboratory. Do not rely on naked eye appearance. **Red cells:** $>10^5/\text{mm}^3$ is positive. **White cells:** $>500/\text{mm}^3$ is positive. A positive result requires exploratory laparotomy. False negatives occur with retroperitoneal haemorrhage.

Complications Haemorrhage; organ perforation \pm peritonitis; infection.

Principal source: *Advanced Trauma Life Support*, American College of Surgeons

Injury to nerves and arteries

Nerve injury classification *Neurapraxia* implies temporary loss of nerve conduction often via ischaemia following pressure (eg to the lateral popliteal nerve as it crosses the neck of the fibula, see below). In mixed nerves, the motor modality is the more vulnerable component. *Axonotmesis* entails damage to the nerve fibre but the endoneural tube is intact, providing guidance to the regrowing nerve. Good recovery is the rule. Growth rate is ~3mm/day. *Neurotmesis* means division of the whole nerve. As there is no guidance from the endoneural tube, regrowing fibrils cause a traumatic neuroma if they are unable to bridge the gap. The current surgical standard is epineural repair with nylon sutures. To span gaps that primary repair cannot bridge without excess tension, nerve cable interfascicular auto-grafts are used. Results of nerve repair are no better than fair, with only 50% of patients regaining useful function. There is much current research regarding drugs, immune system modulators, enhancing factors, and entubulation chambers.

medline:1095111

Median nerve (C5–T1) This is the nerve of grasp. Injury above the cubital fossa causes inability to flex the interphalangeal joint of the index finger on clasping the hands (Ochner's test); inability to flex the terminal phalanx of the thumb (flexor pollicis longus); loss of sensation over the thenar half of the hand.

If the lesion is at the wrist, the only muscle reliably affected is abductor pollicis brevis. Test it by holding the hand palm up. Can the patient raise the thumb out of the plane of the hand? The area of sensory loss is smaller than that for higher lesions.

Ulnar nerve (C8–T1) This is the nerve of finger abduction and adduction (among other roles). Injury distal to the wrist produces the claw hand deformity [flexor digitorum profundus is intact, but is not counteracted by the small muscles of the hand—the basis of Froment's paper sign: on holding a piece of paper between thumb and finger (both hands), there is flexion of the thumb's distal phalanx on trying to pull apart]. Sensory loss is over the little finger, and a variable area of the ring finger.

Radial nerve (C5–T1) This is the nerve of extension of the elbow, wrist, and fingers. It opens the fist. Injury will produce wrist-drop. Test for this with the elbow flexed and the forearm pronated. Sensory loss is variable, always including the dorsal aspect of the root of the thumb.

Sciatic nerve (L4–S2) Complete lesions will affect all muscles below the knee, and sensation below the knee laterally.

Lateral popliteal (common peroneal) nerve (L4–S2) Lesions lead to equinovarus with inability to dorsiflex the foot and toes. Sensory loss is over the dorsum of the foot.

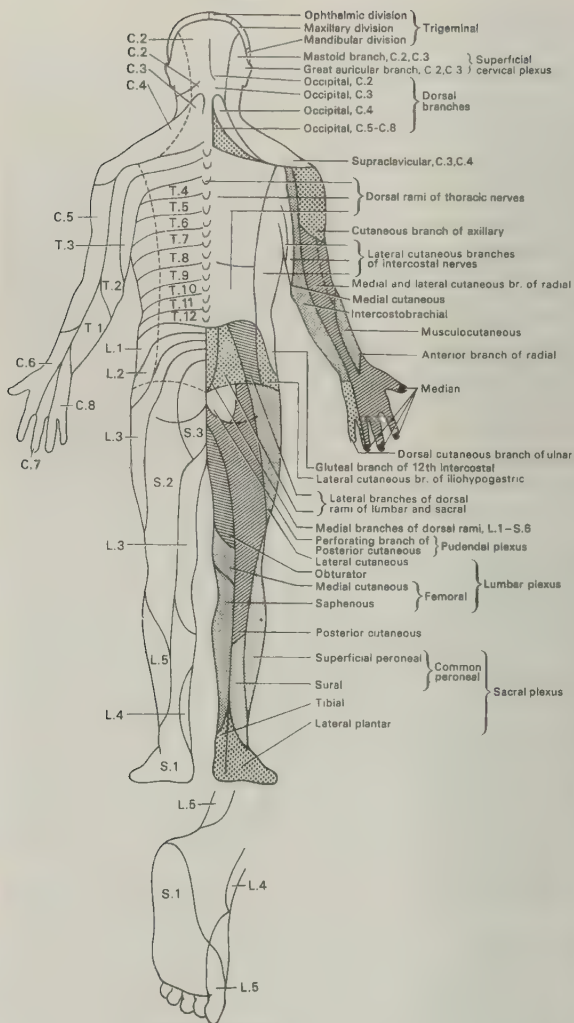
Tibial nerve (S1–3) Loss causes calcaneovalgus and inability to stand on tiptoe or invert the foot. Sensory loss over the sole.

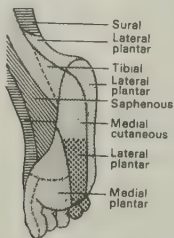
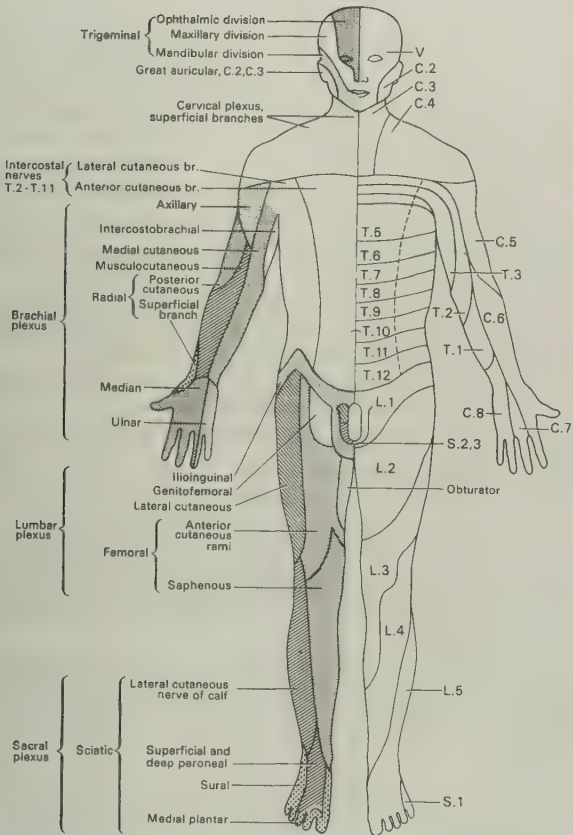
Injuries to arteries Bleeding is usually controllable by pressure and elevation of the part. After any injury in which an artery may have been damaged, examine distal pulses carefully. If they are not felt, do not assume that this is due to spasm, but request expert help. Surgical exploration with end-to-end suture, or reversed vein grafts may be needed. The prognosis is not so bad if there are good collaterals (eg to the femoral artery, compared with the popliteal artery). *Complications:* gangrene, contractures, traumatic aneurysms, and arteriovenous fistulae.

Sensory relearning after median nerve repair: the Lundborg–Rosén metaphor

Tactile gnosis: This is tested by 2-point discrimination and tactile recognition of objects. Restoration of tactile gnosis in the hand is the main challenge of median nerve repair. It is easy to point out that touch is a major way we interact with our world. More subtle is how the brain performs *cross-modal interpretation* of our world—ie integrating touch, sound, and vision, and producing meaning out of this integration. Loss of one modality is not a simple case of subtraction, but gives rise to complex compensatory mechanisms. Interpretation of textures, for example, is not usually regained after major injury to a nerve trunk in adults, but there is more plasticity in children: here, cortical remodelling allows meaningful interpretation from mis-connected regrown axons. Best results of repair are seen before the age of 10yrs old. By 18yrs, there has been a rapid decline in relearning ability. There is an unexplained temporary increase in learning ability in the late twenties. This pattern, intriguingly, follows that of our ability to learn a new language. So the notion that the problem after nerve injury/repair is that '*the hand is speaking a new language to the brain*' is more than an idle metaphor; accepting this metaphor leads to the idea that education and training are vital to successful nerve repair. G Lundborg 2001 *Lancet* 358 809

Dermatomes





ANTERIOR ASPECT

Injuries to the brachial plexus

These may be caused by pressure from crutches to the cords in the axilla; cervical traction and lateral flexion from forceps deliveries or falls from fast-moving motorcycles (eg C5, C6 ± paralysed diaphragm); or tumours, bullets, cervical ribs, or radiation (eg for breast cancer). Complete lesions (C5–T1) produce a flail, anaesthetic arm (except for a medial strip along the arm supplied by the intercostobrachial branch of the second intercostal nerve).

Root injuries—Erb's palsy (C5, C6) There is paralysis of deltoid, supraspinatus, and teres major. Because external rotation of the arm is diminished, and biceps paralysis prevents flexion, it is held internally rotated, and if adducted behind the back it adopts the 'porter's tip' position. Sensation is impaired over deltoid, lateral forearm, and hand.

Klumpke's paralysis (C8, T1) The arm is held in adduction. There is paralysis of the small muscles of the hand, with lack of sensation over the ulnar side of the arm. Horner's syndrome (p 488) may occur.

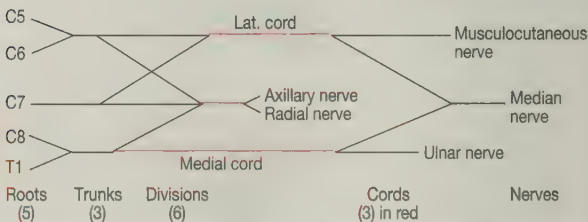
Injury to the cords • Injury to the lateral cord of the plexus: absence of power in the biceps and coracobrachialis (flexes the wrist).

• Injury to the posterior cord: teres major and deltoid inaction, and radial nerve palsy.

• Injury to the medial cord affects the ulnar and median nerves (p 720). Sensation is absent over the medial arm and hand.

Recovery With incomplete trunk lesions any recovery may take >12 months. The prognosis is poor in lesions proximal to the dorsal root ganglion (DRG). For intradermal histamine (1%, into the affected limb) to produce an arterial flare, the route to the DRG must be intact, so if this is present the lesion is proximal to the DRG. MRI gives the best images. Seek expert help early.

Prompt reimplantation of avulsed nerve roots may be attempted with nerve transfers in severe cases of brachial plexus injury. medline:1101205 Tendon transfers, shoulder arthrodesis and other procedures may be indicated. Chart recovery with serial 2-point discrimination tests.



This diagram of the brachial plexus, the *bête noire* of all medical students is included not to give a life-like picture, but to allow a scheme that can be learned the night before an exam. Its only virtue is that it is simple enough to allow you to reproduce it from memory on the back of an examination paper which might be asking you such arcane questions as 'can fibres from C5 contribute to the median nerve?' (answer: yes)—or are there more trunks than divisions (answer: No).

Spinal cord injury—the first day

At the site of the accident Spinal injury should be assumed to be present in any serious accident, and in all accidents when the nature of the injuring force is not known, and the patient is unconscious. This does not mean that the patient cannot be moved into the recovery position (protection of the airway is always the first priority)—but all movements should be planned. Keeping the head in neutral, the patient may be ‘log rolled’ into a semi-prone position (not lateral, see p 789). For immobilization, see p 788.

Radiology Don’t rely on mobile equipment; if possible, take to the radiology department, supervising all movements and the removal of sandbags and collars. Do CT if possible. medline:10405730 With plain radiographs, the lateral view is often the most revealing; do oblique views if the C7–T1 junction is not visible. Anterior vertebral displacement of $< \frac{1}{2}$ the diameter of the vertebral body suggests unilateral facet dislocation. Greater displacements indicate bilateral (unstable) facet dislocations. Look for the soft-tissue shadow of a paravertebral haematoma if bony signs are minimal. *Other views:* anteroposterior (AP) with an open mouth view of the odontoid process \pm AP view of C2–T1 area. Thoracic, lumbar & sacral injuries are usually shown on AP and lateral films. If there is an area you are unsure about, discuss with the radiographer how to show it better. CT is preferred to x-rays of the cervical spine in some dedicated trauma centres. MRI shows fractures, subluxations, disc disruption and protrusion, and cord contusion—and helps establish prognosis.¹

Respiratory insufficiency Check vital capacity repeatedly. If < 500 – 600 mL, intubation and ventilation will be needed. Monitor blood gases. Intubation may produce vagal bradycardia, so give atropine 0.3–0.6 mg iv before intubation and airway suction.

Fluid balance There is likely to be hypotension below the lesion (sympathetic interruption). This is not due to hypovolaemia, and it is dangerous to give large volumes of fluid. Use iv not oral fluids for 48 h and while ileus persists. If abdominal distension causes respiratory embarrassment, pass a nasogastric tube.

The skin Turn every 2 h between supine and right and left lateral positions. The Stoke Manderville bed does this electronically. Use pillows to separate the legs and maintain a lumbar lordosis.

Steroid treatment ▶ Methylprednisolone 30 mg/kg iv stat (within 8 h of trauma) followed by 5.4 mg/kg/h ivi for 23 h is known to significantly improve subsequent recovery;^{1,2} SE: worsening of wound infections.

Early surgical decompression ♦ Get expert advice on removing damaging bone & disc fragments. (Late internal fusions may also be tried.)

Anticoagulation Heparin 5000 u/8 h sc. Start warfarin later.

The bladder Pass a 12-gauge silicone 5 mL balloon catheter before the bladder volume exceeds 500 mL (overstretching of detrusor can delay the return of automatic bladder function). See p 730.

▶ Arrange early and expert transfer to a spinal injuries unit. Skeletal traction will be needed for cervical injuries. Spring-loaded Gardner–Wells skull calipers are preferable to Crutchfield calipers which need incisions.

Does this patient have a spinal cord injury?

In any unexplained trauma, suspect cord injury.

Signs to look for:

- Responds to pain only above clavicle
- Breathing—diaphragmatic
- Muscles—hypotonic, including reduced anal tone (do a PR)
- Reflexes—hyporeflexic
- Slow pulse and ↓BP, but in the presence of normovolaemia
- Priapism

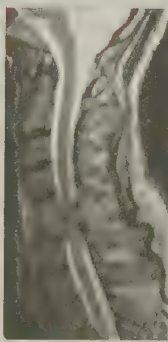
How complete is the injury? **ASIA scale** ASIA=american spinal injury association
A=complete; no sensory or motor function in sacral segments S4–S5.
B=incomplete; sensory but not motor function preserved below the neurological level, and extending through sacral segments S4–S5.
C=incomplete; motor function preserved below the level, most key muscles weaker than grade 3 (ie no movement against gravity).
D=incomplete; motor function preserved below the level; most key muscles are stronger than grade 3, ie active movement against gravity.
E=normal motor and sensory function.

What are the mechanisms of injury?

Primary injury (immediate) is easy to understand: within seconds, the cord expands to occupy the entire diameter of the spinal canal. Glutamate floods out of neurons, overexciting their neighbours. Calcium floods in, leading to the formation of toxic free radicals.

Secondary injury (delayed, eg unfolding over weeks) is a response to release of neurotoxins and apoptosis (cell death) which may spread up to 4 segments away from the trauma site.

www.trauma.org



Because of different technical settings, MRI images can be called T1 or T2 weighted. These images can be recognised as T2 weighted because the CSF is white. Compression of the cord has occurred as a result of a disc protrusion, a metastatic deposit, and trauma.

Cord injury—function and functional anatomy

The 'tightest fit' between cord and canal is in the thoracic spine; this region also has the poorest blood supply. These facts explain why thoracic lesions are more likely to be complete than cervical or lumbar lesions. Ischaemic injury often spreads below the level of the mechanical injury. For the segment of the cord involved with injury at a specific vertebra, see p 608 & p 722.

Cord compression may be due to bone displacement, disc prolapse, local tumour, or abscess. Root pain (p 618) and lower motor neurone signs occur at the level of the lesion with upper motor neurone signs and sensory changes below the lesion (spastic weakness, brisk reflexes, upgoing plantars, loss of co-ordination, joint position sense, vibration sense, temperature and pain). Cord anatomy is such that dorsal column sensibilities (light touch, joint position sense, vibration sense) are affected on the same side as the insult, but spinothalamic tract interruption affects pain and temperature sensation for the opposite side of the body 2–3 dermatome levels lower than the affected sensory level. As the cord ends at L1, compression at this vertebral level affects information in the cord relating to lower dermatomes. To determine the cord level affected behind a given vertebra, add the number in red to that of the vertebra concerned, thus: C2–7: +1, T1–6: +2, T7–9: +3, T10 has L1 and L2 levels behind it; T11 has L3 and 4, L1 has sacral and coccygeal segments. It can be difficult to determine the level: MRI will help clarify this.

Lower lumbar problems can cause cauda equina compression characterized by muscular pain, dermatomal sensory changes (if the lowest sacral dermatomes are affected the genitals are anaesthetic), and retention of urine \pm faeces. ▶ *These signs indicate urgent neurosurgical referral with imaging eg to confirm or exclude a tumour or extradural abscess.*

Spinal shock (BP↓ but no tachycardia.) There is anaesthesia and flaccid paralysis of all segments and muscles innervated below the level affected with areflexia and retention of urine. A 'stage of reorganization' then occurs (reflex emptying of bladder and rectum; sweating). Later, flexion at hip and knee may be induced by stimuli (notably cold), often with emptying of the bladder and rectum (Head and Riddoch's mass reflexes). The legs may become permanently fixed in a drawn up position, with dorsiflexion of the ankles (spastic paraplegia in flexion). Over months, tendon reflexes return, and proprioceptive stimuli give rise to 'mass extension'.

Paraplegia/quadruplegia levels Levels fluctuate, so examine often.

Raises elbow to horizontal: <i>deltoid</i> C5	Flexes hip <i>iliopsoas</i> L2
Flexes forearm: <i>biceps</i> C6	Extends knee <i>quadriceps</i> L3
Extends forearm: <i>triceps</i> C7	Dorsiflexes ankle <i>tibialis anterior</i> L4
Flexes wrist and fingers C8	Plantarflexes ankle <i>gastrocnemius</i> S1
Spreads fingers T1	

Segment injury—a guide to possible subsequent function

- C4 Can use electric chairs with chin control; type with mouthstick; use a 'Possum' environmental control system to turn on lights & open doors.
- C5 With special devices, he can feed, wash face, comb hair, and help with dressing the upper body. He may be able to push a wheelchair along the flat, if pushing gloves are worn, and there are capstan rims on the wheels. The NHS will supply an electric wheelchair (indoor use only). Unable to transfer from wheelchair to toilet.
- C6 Still needs a strap to aid feeding and washing. Dresses top half of body; helps dress lower half. Can drive with hand controls.
- C7 Can transfer, dress, feed himself.
- C8 Independent wheelchair life.

Restoring lost function: is it possible?¹

The following is not a list of well-tried treatment options: the field is too new. The following is more a way of thinking about cord injury, and preparing our minds for discussions with experts. Most patients prioritize their wishes for restoration of functions in this order: bowel and bladder control; sexual function; hand function; breathing. Options might be:

- Glutamate receptor blockers to limit secondary injury (p 726–7).
- Antiapoptotic agents, eg NT-3 (neurotrophin-3), BDNF (brain-derived neurotrophic factor), and ICE-protease (interleukin-1 β -converting enzyme) inhibitors.
- Chemical prevention of action potential dissipation from demyelinated areas \pm agents to remyelinate axons.
- Blocking natural inhibitors of regeneration, eg IN-1 (inhibitor neutralizing antibody).
- Promotion of axonal regeneration—eg BDNF & NT-3, (see above). Note that guidance of axons must be arranged, eg via guidance molecules such as netrins, cell adhesion molecules, and specific matrix proteins.
- Replacement of lost cells (cloned embryonic stem cells are one option; use of endogenous progenitor cells is another option).

Often these options seem a long way off, but note that only 10% of damaged neurones need to be replaced to enable useful locomotion.

¹ J McDonald 2002 *Lancet* 359 417 [@11844532]

Spinal cord injury: genitourinary aspects

One major problem is urinary incontinence and reflex detrusor activity (after acontractility in the period of spinal shock) and the presence of residual urine. This predisposes to infection and ureteric reflux. These are major causes of renal failure, morbidity, and mortality.

The method of bladder drainage In the first weeks after cord injury use suprapubic or intermittent urethral catheterization with a 12 or 14 Fr Nelaton[®] catheter, with strict asepsis. Culture the urine every week and aim to eradicate infection (particularly important with *Proteus* which induces stones in infected alkaline urine). In some patients it may be possible to induce voiding by tapping the suprapubic area for ~20 sec. Initially, catheterization is still necessary to drain the residual urine, but when this is <80mL (on 3 consecutive occasions), discontinue the catheter. If this does not occur, and particularly if the detrusor is non-contractile (conus medullaris or cauda equina injury), intermittent self-catheterization (eg with a silver catheter in women) may be used, as soon as the patient can sit. If reflex voiding occurs, propantheline 15–30mg/8–12h po 1h ac may reduce detrusor activity and obviate the need for wearing drainage devices continuously. The elderly may require an indwelling silicone catheter, with 6-weekly changes. Weekly washouts (eg Suby-G[®]) may prevent stones. Aim for an output of >3 litres/day. Use clamps to achieve volumes of 300mL. Artificial sphincters are available for acontractile bladders.

Complications *Urinary infection:* This may be prevented by a high fluid intake, ensuring effective bladder emptying, and acidification of the urine (eg ascorbic acid 1g/6h po).

Detrusor-sphincter dyssynergia: The external urethral sphincter fails to relax or actively contracts during detrusor contraction. There is poor bladder emptying and vesicoureteric reflux, predisposing to pyelonephritis, hydronephrosis, and renal failure. It is wise to do u&e, creatinine, and ivu as early baseline tests. *Treatment:* Endoscopic external sphincterotomy.

Autonomic dysreflexia: In those with lesions above the sympathetic outflow (ie above T1) bladder distension (eg with a blocked catheter) results in sympathetic overactivity below the level of the lesion. There is vasoconstriction and hypertension (may cause strokes). The patient has a pounding headache, and blotchy skin above the lesion. The carotid baroreceptors are stimulated causing reflex vagal bradycardia, but the signals which would normally produce relieving vasodilatation and normotension are unable to pass down the cord. Other stimuli which may produce this effect include UTI, calculi, labour, and ejaculation. *Treatment:* Remove the cause; give nifedipine (10mg—bite the capsule) with glyceryl trinitrate 0.5mg. Phentolamine 5–10mg iv is an alternative. Sphincterotomy may prevent attacks.

On-demand urination There have been encouraging trials in those with ASIA-A lesions (p 726–7) of the Brindley–Finetech Vocare implantable bladder system which activates anterior sacral nerve roots to regulate bladder and large bowel and urethral/anal contraction. This leads to cost savings (fewer catheters), fewer UTIs, and a better life. medline:11844532

Sexual function

► *Sexual counselling is an integral part of rehabilitation.* It is important in itself, but we should recognize that sexuality interacts with important determinants of our patient's quality of life, eg levels of dependency, aggression, self-esteem, and autonomy. medline:8048654

We should enlist expert help, but we can all counter prevalent myths that disabled men cannot sexually satisfy able-bodied women; and that the cord-injured cannot have intercourse. Don't be shy and don't be shocked: for help with discussing sexual issues, see p 328. ► Given a knowing and patient partner, most persons with spinal injury can enjoy a satisfying sex life. medline:1223314 &: 3604847

We need to be aware that sexuality encompasses more than physical attractiveness and penile-vaginal intercourse. With spinal cord injury, use of sexual imagery and concentration on body areas that retain sensation have especial importance, as does a certain inventiveness and readiness to experiment. medline:6585940

When helping these patients it is important to distinguish sexual drive and sexual satisfaction from fertility and parenting needs. Both need addressing in a systematic way within the broader contexts of psychosocial, emotional, and relationship aspects—and also in terms of cognitive-genital dissociation, perceived sexual disenfranchisement, and sexual rediscovery. Cognitive-genital dissociation may take various forms, eg indicating that for some, sexual activity is more like watching a graphic movie than engaging in a physical experience. medline:11554227
It is equally important to assess the partner's needs and responses to the injury. This takes time. medline:1642822

Do not concentrate on physical aspects alone: nevertheless it may be helpful to familiarize yourself with some statistics, and to emphasize that individuals vary. In some studies, locomotor impairment and dysreflexia were more frequently given as causes of reduced sexual pleasure than specific sexual dysfunctions. medline:6585940

In women, only 17% with complete lower motor neurone dysfunction affecting the S2–S5 spinal segments can achieve orgasm, compared with 59% of women with other levels and degrees of injury.

In men with lesions between T6 and L5, 75% can expect improvement in erections with use of sildenafil. medline:11198294 &: 9855514

Fertility issues in men centre around performance, and sperm quality which may be reduced by scrotal hyperthermia, retrograde ejaculation, prostatic fluid stasis, and testicular denervation. Electro-ejaculation and intracytoplasmic sperm injection have a useful rôle.

In women, pregnancy rates vary from ~10% to 60%. medline:11844532

Cord injury—OT, physio, and nursing aspects

The occupational therapist (OT) is a key person in maximizing the levels of achievement outlined above. She is also in a position to arrange a home visit with a member of the spinal injuries team, and a community liaison nurse or social worker. The aim is to construct a plan with local social services and the local domiciliary occupational therapist, so that the patient's (and his family's) hopes can be realized to the fullest extent. She can arrange the necessary home modifications, and give invaluable professional advice about the level of independence which is realistic to strive for. As ever, the aims of the occupational therapist extend into augmenting self-esteem, and helping the patient come to terms with loss of rôle, and loss of confidence, and to mitigate the effects of disability by arranging for as much purposeful activity as possible, both in the realms of work and leisure. She will also be able to make plans for acquiring of social skills to assist the patient in his or her new way of life.

Nursing & physiotherapy *The chest* Regular physio with coughing and breathing exercises prevents sputum retention and pneumonia which are likely to follow diaphragmatic partial paralysis (eg C3–4 dislocation). If the lesion is above T10 segmental level, there is no effective coughing.

The straight lift: (for transferring patients) One attendant supports the head with both hands under the neck so that the head lies on the arms. 3 lifters standing on the same side insert their arms under the patient, one at a time, starting at the top. After the lift, withdraw in the reverse order.

The log-roll: (Learn from observation.) 3 lifters stand on the same side of the patient. The one near the head has both arms under the patient's trunk and over the patient's further arm; the 2nd has one arm under the legs and the other arm holds the patient's iliac crest. The 3rd lifter supports the calves. A 4th person controls the head and neck, and gives the command to turn. The patient is then gently rolled laterally, with pillows to support the lumbar curve and to keep the position stable.

Posture: Place joints in a full range of positions. Avoid hyperextensions. Keep the feet flexed at 90° with a pillow between soles and bed-end.

Bowels: From the second day of injury gentle manual evacuation using plenty of lubricant is needed. A flatus tube may be helpful in relieving distension once the ileus of spinal shock has passed.

Wheelchairs: The patient should be kept sitting erect; adjust the footplates so that the thighs are supported on the wheelchair cushion and there is no undue pressure on the sacrum. Regular relief of pressure on the sacral and ischial areas is vital. Independence in transferring to bed or toilet will be a suitable aim for some patients with paraplegia. Expert skill is needed in assigning the correct wheelchair for any particular patient.

Standing and walking: Using a 'tilt table', or the Oswestry standing frame with trunk support straps, the tetraplegic patient can gain the upright posture. If the level of injury is at L2–4, below-knee calipers and crutches will enable walking to take place. If the lesion is at T1–8, 'swing to gait' may be possible. The crutches are placed a short distance in front of the feet. By leaning on them and pushing down with the shoulders, both legs may be lifted and moved forwards together.

Sport: Consider archery, darts, snooker, table tennis, and swimming for those with paraplegia. Many other sports may also be suitable.

Personal qualities in therapists are almost as important as exact anatomic lesion. There may be big mood swings from euphoria to despair as the patient accustoms himself to his loss and his new body image.

Vegetative states

"I seemed to have been asleep nearly all my life."
"—But when did you wake up?"
"I don't know that I ever did, or ever have" *Sims & covers 274*

Persistent vegetative state' is a term coined to describe the behaviour of severely brain damaged people who show sleep–awake behaviour, but in whom there is absent cognition and awareness. The term is something of a misnomer, because its presence does not imply permanence (recovery of cognition may occur up to 5yrs after the original insult)—and it has nothing to do with vegetables.

Persistent vegetative states are rare, and liable to misdiagnosis by non-specialist neurologists and neurosurgeons—perhaps because diagnosis depends on history and examination—requiring careful, repeated assessment and the taking into account of observations made by family and carers. ▶*The harder you look, the more likely you are to find signs of active cognition, so negating the diagnosis of a vegetative state.*

One method of showing awareness and cognition is by demonstrating communication, eg by following a simple request to press a buzzer or to look at a named object.¹ These switches are particularly suited to those who can only manage a tiny amount of voluntary movement. Other ways of communicating include listener scanning: the therapist goes through A–Z slowly, and the patient buzzes when the required letter is reached. It may take weeks or months to establish that the patient can communicate. Rancho scale assessment establishes the level of cognition—from confused-and-inappropriate (level 5) to purposeful-and-appropriate (level 8).

Causes Trauma, anoxia, vascular causes, encephalitis.

Management Seek expert rehabilitative help. While waiting for any recovery, aim to provide vigorous nursing care to maintain nutrition, and to prevent pressure sores, and tracheostomy and muscle contracture complications. No drugs are known to help, although there is anecdotal evidence that bromocriptine may help.² Randomized trials indicate that multimodal stimulation (arousal programmes) help.³ These programmes involve stimulation of all senses every 15min for up to 11h per day.²

Once it is agreed that there will be no recovery (a difficult issue), discussions about withdrawing active treatment and nutrition raise difficult ethical questions—which are only partly mitigated if the patient has an advance directive (an unlikely event). Ethicists, medical specialists, nurses, physiotherapists, judges, and the family must all be allowed to have their say. None is infallible.

Does he need a radiograph?

The aim is to provide information that will alter management, without exposing patients to unnecessary radiation. The following is based on the advice of the UK Royal College of Radiologists, and other bodies.

Presenting conditions *Twisting injury of foot or ankle:* A radiograph is indicated if patients cannot bear weight; one is said *not* to be indicated if he can weight-bear on the affected ankle, and if tenderness and swelling are confined to a ligament, or are absent—but all rules have exceptions, and, on occasion, we've all seen patients walk on a fractured ankle.*

Crush injury to the terminal phalanx of a digit: A radiograph is not indicated in localized closed injury, provided there is pain-free movement at the distal interphalangeal joint.

Injury to the cervical spine: If any of these are present, imaging is needed:

- Neurological exam reveals a focal deficit.
- Spine exam reveals tenderness (posterior midline).
- Alteration in consciousness.
- Intoxication.
- Distracting forces as the mechanism of injury.

The Canadian CT head rule: CT is only required for minor head injury if one of the following is present:^{2,3}

- Glasgow coma scale <15 at 2h post-injury.
- Suspected open or depressed skull fracture.
- Any sign of basal skull fracture (haematotympanum, racoon eyes, CSF otorrhoea/rhinorrhoea, Battle's sign—see p 714).
- Amnesia before impact of >30min.
- Dangerous mechanism (pedestrian struck by car; car occupant ejected, fall from a height of >1m, or downstairs).

'Possible' neck injury in minor head injury: A radiograph is not indicated if fully conscious and there are no symptoms or signs related to the neck.

Nose injury: A radiograph is not indicated in the Accident and Emergency department in simple nasal injury.

Rib injury: Only a posteroanterior CXR is indicated if you suspect a pneumothorax; rib views are not needed in uncomplicated blunt injury.

Lumbar spine pain: Avoid early radiographs (in first 6 weeks) if there are no CNS signs or factors suggesting serious disease, eg trauma, fever, weight loss, anaemia, ESR raised (so do blood tests first). Each examination = 2.2mSv (= 40 chest radiographs; the sievert is the SI unit of radiation absorbed by biological tissues—its abbreviation is Sv), which may be expected to cause 16 malignancies/yr in the UK at current rates of exposure. It is impossible to protect the ovaries.

Abdominal pain: Plain films are rarely needed; request *only* an erect chest film in suspected gastrointestinal perforation. Erect abdominal films are generally not indicated in most patients with abdominal pain.

Foreign bodies: Always do radiography if the presence of glass is possible (glass is usually radiopaque).

Note: Lumbar spine radiographs deliver a significant dose of radiation to the gonads—similarly for skull and facial radiographs to the eyes.

These guidelines yield substantial savings in costs and in patients' waiting times, without compromising patient care.

*Ottawa rules, see www.ohri.ca/programs/clinical_epidemiology/OHDEC/ankle_rule/default.asp

Appropriate use of Emergency Departments

From our point of view, the ideal scenario is to have a pristine, ever-ready but empty trauma department with the doctors and nurses educating themselves (etcetera) in the coffee room, occasionally being called out by paramedics to save a few lives in a brief display of energy and technical brilliance. This is not how the public sees our rôle. If you are in shock because you have just run over a hedgehog some people feel they need attention *now*. All Emergency Departments are abused—because it is always doctors who define what abuse is. Thus up to 70% of users have been deemed inappropriate in some studies.

Overcrowding & 'exit-block'¹ This is a major problem in Emergency Departments the world over—partly reflecting centrally-determined reductions in acute bed provision and availability, and partly reflecting access problems for populations seeking prompt help with immediate (and, sometimes, chronic) problems. If the inpatient side of a hospital is full, patients will stack up in Emergency Departments, awaiting admission. If overcrowding is cumulative, ambulances may be redirected to other hospitals (adding to delay and danger) and patients with genuine needs may, catastrophically, decide they cannot wait any more hours for help.

How to cope with inappropriate attendance

- **Triage** by a trained nurse is one way to reduce inappropriate attendance. (In the UK, triage is mandatory.) Is this condition *life-threatening*, *urgent*, *semi-urgent* or is *delay acceptable*? These are the chief categories. If the most urgent patients are seen first, patients who do not really need to be in the department can wait for ages, and many will begin to drift away. Note that if long waits become essential because of dealing with life-threatening conditions, this should be explained to patients (under the terms of the UK health charter—and as a matter of common courtesy).
- Another way to reduce unnecessary spending is to have primary care facilities within the Emergency Department, or, more radically, to have GPs as the casualty officers—because they use resources more sparingly and are used to dealing with mismatches between patient's expectations and reality. Is this expectation borne out in practice? In one careful randomized study of patient-contacts with the Emergency Department in Dublin ($N=4684$), GPs treating people with semi-urgent problems investigated less (by 20%) referred to other hospital services less (by 39%), admitted fewer patients (45%) and prescribed more often (43%): there were no differences in measures of outcome. It is not clear whether the more economical style of the GPs was to do with being a GP, or whether the reason was that these doctors were older and more experienced than their emergency department counterparts.

735

Risk management strategy when overwhelmed by the work to be done:

- Prioritize the waiting tasks and then concentrate on the task in hand.
- At times you may feel you are working at a pace that is faster than is comfortable. This is part of 'continuous learning', and as you get better this unease will abate. Look on it as practice for emergency situations where quick decisions are vital. Working faster does not necessarily mean making more errors.
- Have good techniques of clinical decision making, which you continue to refine. Start with something like the system on p 472.
- Keep the goal in mind *What to do for the best for this patient.*

Bites, stings and foreign bodies

Mammal bites All animal bites are contaminated, especially those of human animals: *everything that comes out of the human mouth is poison*.¹ Clean well with soap and water, debriding if needed. See p 704. ▶ Avoid suturing unless cosmetically essential. Give antibiotics covering anaerobes unless very trivial and not high-risk (risk↑ if: ♀, >50yrs, asplenic, immunosuppressed/compromised, has alcoholic liver disease, or the wound is to hand or foot), eg co-amoxiclav 1tab/8h po (clindamycin 300mg/6h po if allergic to penicillin but beware pseudomembranous colitis). NB: bites from cats may not be as trivial as they look: they carry *Pasteurella multocida*, streps, and fusibacteria. Is tetanus prophylaxis needed? Consider rabies if bitten outside uk. (Post-exposure rabies prophylaxis: phone the uk Central Public Health Lab, 0181 200 4000.)

Snake bites Britain's only poisonous snake, the adder, is very rarely lethal. Identify the species if possible and move the patient to hospital at once. Treat shock, monitor clotting time (many venoms are anticoagulant), and watch for renal failure, respiratory paralysis, and tissue necrosis. Indications for antivenom treatment include systemic envenoming, as signalled by hypotension, wcc raised—as well as local envenoming which is spreading beyond the wrist and ankle more than 4h after the bite (for bites in these locations). Give European viper venom antiserum (see *BNF*) 1 vial iv over 15 mins. Use the same dose for children. It may be repeated after 1h, if there is no improvement. Have epinephrine (=adrenaline) to hand (p 309). If the bite is from a foreign snake or spider, the relevant antivenom may be found at Fazakerley Hospital Pharmacy, Liverpool (0151 525 5980). Information on identification and treatment is available.² ▶ Tourniquets, incisions, and sucking the wound must be avoided (but a bandage to the limb may help confine venom).

Lesser weever fish stings (*Echiichthys vipera*)—eg in barefoot uk bathers. It is not serious, and may be relieved by immersion of the leg for 5–20min in water which is as hot as can be tolerated (eg <45°C).

Scorpion bites Prazosin helps; lidocaine sc at the site relieves pain.

Bee stings Scrape out with a knife or credit card *quickly*. Pheromones released from the expiring bee will attract more bees. You may well be able to outrun them if you see them coming. NB: although always fatal to the bee, a single sting almost never kills you (risk <1 in 1,000,000)—but fatalities are likely if you receive >2000 stings. Ice ± calamine lotion help itch. Give antihistamines for severe swelling/itch. Anaphylaxis, p 309.

Airgun pellets These are common, and can be hard to remove. Deaths have occurred, eg when a pellet enters brain through the eye. Get 2 radiographic views to position the foreign body. If it has just penetrated the skin, inject local anaesthetic carefully so that you can still palpate it: if you cannot remove it easily, leave it *in situ* rather than risk extensive tissue destruction trying to find it; give antibiotics. Pellets tracking subcutaneously, or which have penetrated deeply, must be sought.

Other foreign bodies ▶ Always do radiography if there may be glass in a wound. Tiny shards may be left *in situ*. Even large shards can be hard to find, needing exploration under GA to remove (also true for needles/pins stuck in the sole). ▶ Always do orbit radiography for a high-velocity metallic foreign body, that cannot be seen (eg grinding/hammering injury).

Heimlich manoeuvre and back blows See p 785.

¹ John Steinbeck 1945 *Cannery Row*, Minerva, 410 ² Oxford 01865 220968/741166, Liverpool 0151 708 9393, London 0207 1 635 9191

Coin ingestion

If symptomatic, arrange chest & neck radiograph (AP and lateral, to differentiate GI and respiratory tract location). Abdominal films may be needed. If below diaphragm, let it pass. If in upper $\frac{1}{3}$ of oesophagus, remove it with an endoscope. Make sure that there are no more foreign bodies before removing the scope; if lower, repeat radiography after 12–24h. If the coin is still in the oesophagus, remove endoscopically under GA.

Common minor procedures

► When in doubt, ask the trauma nurse: she will have seen it all before.

'I've hammered my finger, doctor' This usually causes a subungual haematoma—relieved by expressing the blood through a hole trephined in the nail, using a 19G needle. No force is needed: simply twiddle the needle vertically on the nail: cutting edge will make a suitable hole (p 703).

'I've swallowed a fish bone and it's stuck' Always examine the throat and tonsils carefully. Often the bone has only grazed the mucosa. Use a good light, and grip the tongue with gauze to move it out of the way before removing any visible bones with forceps. If you fail, refer to ENT.

'My fish hook has barbed my finger' Infiltrate with plain lidocaine (=lignocaine), and push the hook on through the finger—provided no important structures are in its path. Once the barb is through, cut it off. Remove the hook by the way it entered.

'My tooth has been knocked out' Try to replace permanent teeth. Send deciduous teeth to the tooth fairy. If the former, after the patient sucks it clean (do not use water) transport in milk—or reinsert it, stabilizing with finger pressure (or biting). Go to a dentist for splinting.

Plaster 'back slabs' (for undisplaced forearm fractures). ²

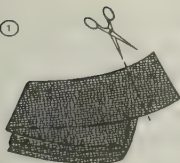
- Remove anything which impairs finger circulation (eg rings).
- Protect yourself and your patient with a plastic apron.
- Measure the length for the back slab—from knuckles to just below elbow, so that the fingers and elbow will remain mobile.
- Cut a piece of plaster-impregnated bandage 5 times longer than the desired length. Fold it into 5-ply. (Fig ①, OPPOSITE)
- Cut off one corner so that it does not impinge on the thumb.
- Cut a wedge off the other end—the wedge's thick end being on the same side as the thumb. This aids elbow movement. ②
- Roll stockinette over the forearm, to well above the elbow.
- Wind a roll of wool padding over the stockinette (turns must overlap by 50%, so protecting flesh from the hard plaster). ③
- Immerse the plaster bandage in tepid water and apply it to the dorsum of the arm—without pitting it with your finger tips. ④
- Reflect the stockinette down from the elbow and up from the wrist making comfortable top and bottom ends to the plaster. ⑤
- Place a bandage right around the forearm to keep everything in place, securing its end with a strip of wet plaster). ⑥
- Setting takes place over 4min: sooner if warm water is used.
- Put the arm in a sling for 1 day—after which encourage movement of shoulder, elbow, and fingers to prevent stiffness.

► **Cautions for the patient:** 1 Return immediately to A&E if the fingers go blue, swell, or you cannot move them. 2 Do not get the plaster wet. 3 Do not lift heavy weights with the hand.

How to remove a tight ring from a swollen finger Wind some cotton tape around the finger, advancing towards the ring. When it is reached, thread the tape through it. Grasp this end of the tape and unwind. This levers the ring over the PIP joint. If not, try using a ring cutter (not for brass or steel).

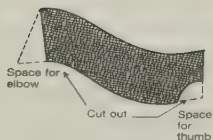
'I've caught my penis in my zip, doctor' Failing simple measures (copious lubrication with mineral oil), the most elegant solution to this is to cut out the bridge from the slider of the zip with strong wire-cutters as shown in the diagrams. The zip then falls apart and all that is needed is a new zip. (Beware the bridge flying off at speed: hold gauze by it.) ²

1

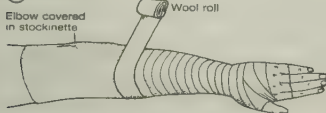


Plaster bandage folded into 5-ply

2



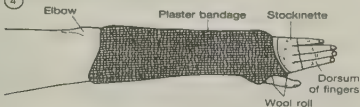
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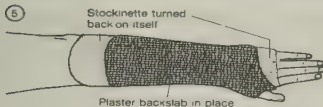
Elbow covered in stockinette

Wool roll

4



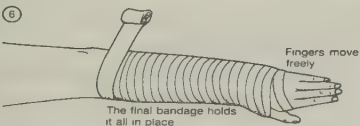
5



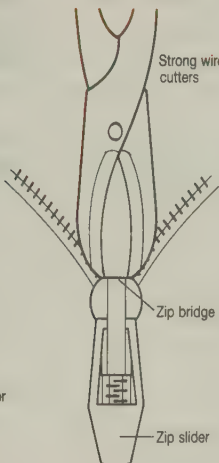
Stockinette turned back on itself

Plaster backslab in place

6

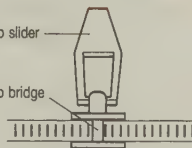


Strong wire cutters



Zip slider

Zip bridge



Top view



Zip bridge

Zip slider

Side view

Heat exhaustion/heat injury

This occurs when core body temperature rises eg to $>39^{\circ}\text{C}$. Basal metabolic rate produces 100kcal/h; radiation from the sun can contribute 300kcal/h, and strenuous muscular activity 900kcal/h (enough to raise the core temperature by 1°C every 5min). Cooling is achieved mainly by evaporation. Above environmental temperatures of 35°C , 75% of heat loss is by sweating. Convection usually accounts for 15% of heat loss, more if windy, but if the environment is very hot it can contribute to heat gain. Acclimatization to a hot environment (metabolic and cardiovascular changes reduce body core temperature and reduce the threshold for sweating), are mainly completed by 7–12 days.

Those at risk from heat illness are those who are: elderly; obese (reduced cardiovascular reserves); children (reduced sweating); exercising in the heat (eg soldiers); with acute febrile illness; on drugs (cocaine, ecstasy, lysergic acid diethylamide—LSD, tricyclics, amphetamines); or dehydrated.

Forms of heat illness

- Oedema: swelling of hands, feet, and ankles in the first days of exposure (worse after long-haul flights). It settles spontaneously over 28 days. Avoid diuretics.
- Heat cramps: painful large muscle cramps after a lot of sweating. Treat with oral rehydration (half teaspoonful salt to each litre of water).
- Heat tetany: hyperventilation induced by rapid change in temperature. Treat as for usual hyperventilation.
- Heat syncope.
- Heat exhaustion: weakness, fatigue, light headedness, nausea, vomiting, cramps, pilo-erection. Core temperature is $<40^{\circ}\text{C}$. Treat with copious fluids eg 1L/h po; and cooling with moist spray and increased convection (fan). Rarely, iv fluids are needed.
- Heat stroke: this is acute neurological impairment with core temperature $\geq 40^{\circ}\text{C}$. Tachycardia, tachypnoea, hypotension, irritability, confusion, seizures (\pm hyponatraemia), and coma may occur. Skin may be hot and dry. Mortality: $\sim 10\%$. Complications include persistent neurological damage, congestive cardiac failure, centrilobular liver necrosis (which can lead to liver failure), acute renal failure (especially in those in whom exercise contributes) due to rhabdomyolysis. Haematuria and proteinuria feature early, DIC can occur at 12–36h. Cool as above. Cold gastric lavage and peritoneal lavage are sometimes used to increase cooling.

medline:10530529

Prevention of heat illness: Avoid exercise in extreme heat, and wear loose fitting clothing only. Keep well hydrated—especially if exercising; eg 500mL po prior to exercising and 200mL/15min whilst exercising.

Electrocution

This accounts for 1000 deaths per year in the USA. Lightning strikes kill between 150 and 300 people per year and seriously injure 1000–1500. Utility workers and those working with electricity lines are the most commonly affected. Injuries within the home are usually due to using improperly earthed appliances or using electrical appliances near water. Alternating current is more dangerous than direct current of the same magnitude.

- Ensure the source of current is turned off before attempting rescue.
- Tetanic muscle spasm may make it impossible for the victim to let go of the source.
- Use paradoxical triage: resuscitate the apparently 'dead' before attending to the living. Using standard resuscitation techniques, resuscitate long and aggressively—remarkable recoveries have been reported.
- Assume blunt trauma (stabilize the spine).
- If there are burns, more fluid replacement is required than for normal thermal burns (7mL/kg/% rather than the standard 4mL/kg/%).
- Treat arrhythmias conventionally.
- Nerves preferentially conduct electrical current resulting in preferential damage to them when other tissues remain intact.

10. Unusual eponymous syndromes

To have any disease is unfortunate, but to have a rare disease is doubly so: the patient must often wait for ages for a diagnosis, and then he must contend with his physician's lifelong morbid interest in him.

For syndromes relevant to *general medicine* and *surgery*, see *OHCM* p 720.

Alport's syndrome X-linked or autosomal recessive sensorineural deafness + pyelonephritis, haematuria, & renal failure (glomerulonephritis + basket weaving of basement membrane). The X-linked forms are caused by mutations in *COL4A5* genes that encode the $\alpha 5$ -chain of type-IV collagen. Typical age at death (σ): 20–30yrs. Medline 2001
M11465475

Asperger's syndrome Autistic features (p 392) *without* autistic alone-ness and linguistic difficulties. It carries a better prognosis than autism.

Bardet-Biedl syndrome Retinal dystrophy, IQ↓, hypogonadism, obesity (\pm polydactyly) + renal abnormalities (calyceal clubbing or blunting, calyceal cysts or diverticula, fetal lobulation). Absence of CNS signs (paraparesis) distinguishes it from the Laurence–Moon–Biedl syndrome (which is better named Laurence–Moon syndrome, p 752). $\sigma/\phi = 47:41$. Genetic locus: 16q13–q21 (recessive). A high incidence is found in Newfoundland (1:17,500 vs 1:160,000 elsewhere). Medline 1998
M9782090 Medline 2001
M11673413

Batten's syndrome (Juvenile neuronal ceroid lipofuscinoses) ↑Apoptosis of photoreceptors & neurones from defects in the *CLN3* gene causes vision↓, childhood dementia, fits, ataxia, spasticity, athetosis, dystonia, and early death (in teens). **Tests:** Lipopigments in lymphocytes & urine. Medline 2002
M11830536

Becker's syndrome This sex-linked recessive muscular dystrophy has a slower progression than Duchenne's, but late onset + more calf enlargement in adolescence. Heart involvement may occur (eg in 50%). **Cause:** Eg lysine to glutamine dystrophin gene mutation (Xp-21 defects). **Treatment:** A few respond to steroids: try and see (under expert guidance; monitor plasma creatine kinase). Medline 2001
M11732779 Medline 2001
M11694896

Beckwith–Wiedemann syndrome Large tongue and kidneys, hemihypertrophy, microcephaly, hypoglycaemia, and omphalocele, plus early feeding difficulties. Risk of neoplasia: 8%: half of these are Wilms' tumours (p 220). Genes involved are at the 11p15 locus; their dysregulation can allow too much insulin-like growth factor 2 to be made. Medline 2001
M9078193

Bourneville's disease (Tuberous sclerosis; epiloia) Multi-organ calcified hamartomatous tubers—autosomal dominant, loci on 9q34 (*TSC1*, making 'hamartin') & 16p13 (*TSC2* makes 'tuberin'; mutations here are most serious).

CNS: Calcified cerebral tubers (≤ 2 cm; may transform to gliomas), \pm seizures, hydrocephalus, periventricular calcification, and a low IQ.

Skin: Hypomelanotic macules (Wood's lamp +ve), adenoma sebaceum (warty nasolabial angiofibromas), *café-au-lait* spots, butterfly rash, periungual fibromas, sacral plaques (Shagreen patches, like shark skin), skin tags.

Mouth: Pitted tooth enamel, gingival fibromas.

Eye: Phakomata (white or yellow retinal tumours; present in ~50%).

Bone: Sclerotic lesions, phalangeal cysts, hypertelorism.

Kidney: Polycystic enlargement, haematuria. **Hair:** White lock.

Lungs: Honeycomb lung, pneumothorax, 'muscular hyperplasia of lung'.

Heart: BP↑, cardiomyopathy, rhabdomyomas, haemopericardium.

Lymph nodes: Castleman tumour. **Bowel:** Polyps \pm Peutz–Jagers' *OHCM* p 732.

Liver & peritoneum: Carney's complex (p 647). **Also:** Precocious puberty.

Tests: Fundoscopy, skull views, CT/MRI, EEG, heart and renal ultrasound.

Prognosis: Often benign; malignancy is rare. www.bmj.com/cgi/content/full/318/7190/1019

When entertaining the diagnosis of a rare disease...

- Ask whether other members of the family have been affected.
- Draw up a family pedigree (as per *OHCM* p 33).
- Have there been unexplained miscarriages or stillbirths in the family?
- Who is now pregnant (or might be, or become so) in the family? In your hands you may hold vital information or vital clues which need to get to the right person at the right time: and that person may not be the person who is in front of you now. Get consent from your patient to talk to the whole family.
- Refer for genetic counselling—as per p 212.
- Is prenatal diagnosis available? If this is a critical issue, don't rely on the judgment of a single expert. Keep on asking, as the field of prenatal tests is constantly enlarging.
- Once a diagnosis is made, don't assume that the disease is untreatable today just because it was last year. Search Medline (p 447) and find out what's changed. Get in touch with a patient's group.

Briquet's syndrome (Fat file syndrome) Hypochondria (p 334) + somatization with >13 unexplained symptoms in various organs.

Bruton's sex-linked agammaglobulinaemia (♂ only) Severe infections and enlarged joints. Peyer's patches, tonsils, and appendix are absent. *Cause*: One of 463 mutations in tyrosine kinase gene. Medline 2001; 11892085

Buchanan's syndrome A single artery arises out of the base of the heart, supplying pulmonary and systemic vasculature. The aorta may be divided. There is cyanosis from birth. Surgical correction is possible.

Capgras' syndrome A delusion that a friend has been replaced by an exact double. *Cause*: Psychosis; head injury; cerebral atrophy; watershed cerebral infarct; right occipitotemporal lesions; pituitary tumour. Medline 2001; 11511338

Castleman's disease Angiofollicular lymph node hyperplasia + benign vascular mediastinal tumour. **2 types**: Hyaline vascular and plasma cell type (causes fever, anaemia, weight loss). Development of frank lymphoma is rare. POEMS syndrome may be present (OPPOSITE). Medline 2002; 11683346

Chediak-Higashi syndrome Immunodeficiency ± albinism, photophobia, nystagmus on exposure to light, weakness, tremor, fever, platelets ↓, liver ↑, ± lymphoma. WBCs contain big peroxidase granules. *Cause*: 1q43 mutation. Fatal in 90% by 10yrs of age without marrow transplantation. Medline 2002; 11857544

Conradi-Hünemann syndrome (Chondrodysplasia punctata) Saddle nose, nasal hypoplasia, frontal bossing, short stature, stippled epiphyses, optic atrophy, cataracts, IQ ↓, flexural contractures. *Cause*: Genetic or effects of warfarin given during the 1st trimester of pregnancy. Medline 2001; 11701653

Cornelia De Lange syndrome Short stature + abnormally shaped head with brows meeting in the midline ± hirsutism, low-set ears, widely spaced teeth, Simian (single) palmar crease, IQ ↓, seizures, self-harm, abnormal temporal lobes. Autosomal dominant, sporadic in 99%. Medline 2002; 1185332

Corrigan's syndrome This is congenital aortic regurgitation.

Cotard's syndrome The patient denies his existence (nihilistic delusion) or he believes that his bowels are rotting, or requests burial, thinking that he is a corpse. According to Camus, 'there is in every man a deep instinct which is neither that of destruction, nor that of creation. It is simply the longing to resemble nothing'. *Causes of nihilism*: Depression, alcoholism, and rarely syphilis or a parietal lobe lesion. Medline 2000; 10965550

Crigler-Najjar syndrome Jaundice ± CNS signs appear in the 1st days of life due to ↓glucosyl transferase activity (?from homozygosity for G71R & Y486D genes). *Rx*: Liver transplant before irreversible kernicterus. Medline 2002; 11792980

Dandy-Walker syndrome Congenital obstruction of the foramina of Luschka and Magendi leads to progressive enlargement of the head, congested scalp veins, bulging fontanelle, separation of the cranial sutures, papilloedema, bradycardia. *Treatment*: Drain the CSF into a body cavity.

De Clerambault's syndrome (Erotomania) The patient is deluded that someone of a higher social status is in love with her. She derives satisfaction from having been 'chosen', and may make trouble by publicizing her view of his feelings. ▶ *Always have a chaperone!* Homoerotomania may be complicated by Fragoli delusions (eg the love object of a teenage boy was a neighbour believed by him to be his father). Stalking is one manifestation. *Associations*: IQ ↓; schizophrenia; mania; left frontal lobe lesion. Medline 1996; 8809668

Diamond-Blackfan syndrome (Erythroblastosis imperfecta) ↓Erythroid production (Hb ↓, platelets ↑, MCV ↑) causes pallor ± limb anomalies. Steroids ± marrow transplant are tried. *Cause*: ?sporadic deletions on 19q13. Medline 2002; 11931266

Helping people with Briquet's syndrome

- Give time—don't dismiss these patients as "just the 'worried well'".
- Explore with the patient the factors perpetuating the illness (disordered physiology, misinformation, unfounded fears, misinterpretation of sensations, unhelpful 'coping' behaviour, social stressors).
- Agree a management plan which focuses on each issue and makes sense to the patient's holistic view of him or herself.
- Treat any underlying depression (p 342); consider cognitive therapy.

Castleman's lymph node hyperplasia with POEMS syndrome

POEMS syndrome entails: **p**eripheral neuropathy, **o**rganomegaly/hyperplasia, **e**ndocrinopathy, a **m**onoclonal paraprotein, and **s**kin lesions. Interleukin-6 excess is also a feature of Castleman's disease, so also look for **p**uo and chronic inflammatory symptoms \pm failure to thrive. Children with unexplained chronic inflammatory symptoms may need detailed soft tissue tests to reveal associated vascular tumours.

Di George's syndrome Absent thymus, small parathyroids, anaemia, lymphopenia, hypocalcaemia, cell-mediated immune deficiency, and seizures. Most are associated with a monosomy for a region of chromosome 22q11. It is related to the velo-cardiofacial syndrome of multiple anomalies, cleft palate, heart defects, cognitive defects, and a characteristic face. Medline 1999 11720799

Di Guglielmo's disease (erythromyelosis) Dysplastic RBCs infiltrate liver, spleen & heart. Hb↓; MCV↔; WCC↓, platelets↓; LDH & B₁₂↑. Immunoperoxidase stains show antihaemoglobin antibody. Transfusions are needed. Medline 1999 1035473

Duchenne's muscular dystrophy presents in boys of 1–6yrs, with a waddling, clumsy gait. On standing, he uses his hands to climb up his legs (Gower's manoeuvre). Distal girdle muscles are affected late; selective wasting causes calf pseudohypertrophy. Later wheelchairs are needed (eg at 9–12yrs). Scoliosis and many chest infections occur. Aim to keep the boy walking (eg by using knee–ankle–foot orthoses). Spinal fixation (Luque operation) or bracing helps scoliosis. Prednisolone 0.75mg/kg/24h helps, but doesn't allow wheelchair abandoning. There is progressive hypoventilation: the cause of death in ~70%. Vital capacity of <700mL is a bad sign. Extending life by ventilation is unlikely to increase its quality. **Creatine kinase** is ↑; measure this in all boys who are not walking by 1½yrs, so that genetic advice may be given (sex-linked recessive). **Muscle biopsy**: Abnormal fibres surrounded by fat & fibrous tissue. **Mortality**: 75% are dead by 20yrs; 5% are alive after 50yrs. ~80% of carrier ♀ have abnormal chemistry. Prenatal screening is available.

Ebstein's anomaly A congenital defect with downward displacement of the tricuspid valve (± deformed leaflets) atrializing the right ventricle. There may be no symptoms, or cyanosis, clubbing, triple rhythm, systolic, and diastolic murmurs. It is to be associated with other cardiac malformations.

Tests: ECG: tall P waves, ↑P–R interval; right bundle branch block. Medline 2001 1763598

Edwards syndrome Trisomy 18, our 2nd commonest trisomy; ♀:♂=2:1. Signs: rigid baby + limb flexion, odd low-set ears, receding chin, proptosis, rocker-bottom feet, cleft lip/palate, ± umbilical/inguinal hernia; short sternum makes nipples look widely separated. Fingers cannot be extended; the index finger overlaps the 3rd digit. Girls survive longer (mean 10 months). Medline 1999 10480700

Ehlers–Danlos syndrome (EDS) Bennett's paradox: the woman at a drag ball is the true imposter for, unlike everyone else, she is what she seems. So with EDS which does not behave like a connective tissue disease because it really is one (a true disease of collagen). Other connective tissue diseases are really diseases of something else. EDS causes soft, hyperelastic, poor-healing, easily bruising skin ± aneurysms or GI bleeds/perforation + loose, hypermobile joints ± flat feet. There are 6 types. Type II, eg, is caused by COL5A mutations, while in type IV COL3A1 mutations upset encoding of type III collagen.

Eisenmenger's syndrome A congenital heart defect which is at first associated with a left to right shunt may lead to pulmonary hypertension and shunt reversal. If so, cyanosis develops (± heart failure and chest infections), and Eisenmenger's syndrome is present. **Prognosis**: poor. Medline 2002 11834351

Erb's scapulohumeral dystrophy ♂:♀ ≈ 1:1. **Onset**: 1–6 decades. **Early sign**: Cannot raise hands above head, then (in order): deltoid→erector spinae→trunk muscles→pelvic girdle→thigh muscles. Lifespan is shortened. Medline 1999 1332322

Fallot's tetrad Pulmonary stenosis, overriding aorta, interventricular defect, RVH. **Signs** Cyanosis as ductus closes, dyspnoea, faints, squatting at play, clubbing, thrills, absent pulmonary part of S₂, harsh systolic murmur at left sternal base. **Tests** FBC: Hb↑. CXR: wooden shoe heart contour + RVH. ECG: RVH; MRI. **Prognosis** 86% 32yrs survival after corrective surgery. 'Total repair' entails VSD closure and removal of the pulmonary stenosis: it may result in normal life, with driving possible, if no syncopal attacks.

X-linked muscular dystrophies

- 1 Duchenne's muscular dystrophy (severe)
- 2 Becker muscular dystrophy (benign)
- 3 Emery–Dreifuss muscular dystrophy (benign; early contractures)
- 4 McLoed syndrome (benign with acanthocytes)
- 5 Scapuloperoneal (rare).

Myotonic dystrophy & other autosomal muscular dystrophies

- 1 Myotonic dystrophy (autosomal dominant; Steinert disease)
- 2 Congenital myotonic dystrophy
- 3 Facioscapulohumeral muscular dystrophy (Landouzy–Dejerine)
- 4 Autosomal recessive Duchenne-like limb-girdle dystrophy of early childhood
- 5 Late-onset (Erb-type) autosomal recessive limb-girdle dystrophy (usually scapulohumeral; rarely pelvifemoral) OTM 3e 4145
- 6 Autosomal dominant limb-girdle dystrophy
- 7 Oculopharyngeal muscular dystrophy
- 8 Distal myopathies
- 9 Non-progressive myopathies. www.homepages.hetnet.nl/~b1beukema/ziekspieren.html

Post-op problems: •Residual vsd + pulmonary hypertension, if big •Ven-tricular tachycardia •Complete heart block •Right ventricular aneurysm •IE/SBE (risk is quite low) •Dilated cardiomyopathy •Pulmonary or aortic regurgitation. NB: when pregnant (may be unproblematic), do careful fetal echoes. Medline 2001 M11752982

Fanconi anaemia Autosomal recessive progressive marrow failure + absence of radii, thumb hypoplasia, syndactyly, missing carpal bones, skin pigmentation, microsomia, microcephaly, strabismus, cryptorchidism, IQ↓, deafness, short stature, and ↑risk of malignancy. *Survival:* 70% at 5yrs post-marrow transplant (umbilical cord blood stem cells are also tried). There are 8 groups: FA-A to FA-H. FAA and FAC genes are the commonest. Medline 2000 M11530803

Galeazzi fracture Radius shaft fracture with distal ulna subluxation.

Ganser syndrome Disorientation plus pseudodementia with 'approximate answering', eg in answer to 'What is the colour of the chair in the corner?' it might be answered: 'What corner? I don't know what a corner is. I don't see a chair. . .'. Absurd remarks only occur as answers to questions. Intellectual deficit is inconstant (hence the 'pseudo'). Hysteria, hallucinations, and fluctuating consciousness are common. It may follow head injury. Medline 2000 M11743582

Gaucher's syndrome (autosomal recessive) The glucocerebrosidase gene is on chromosome 1q21; mutations here cause Gaucher's, the commonest lysosomal storage disease. Glucosylceramide accumulates in macrophage lysosomes (producing Gaucher's cells), causing face & leg pigmentation, and progressive organomegaly, with marrow & CNS infiltration. *Diagnosis:* Skin fibroblast glucocerebrosidase assay. Death may be from pneumonia or bleeding. *Other signs (infantile form):* rigid neck, dysphagia, catatonia, hyper-reflexia, IQ↓. *Treatment:* Imiglucerase may induce remission; escalating doses may be needed (eg from 20–60u/kg/2 weeks ivi to 100u/kg/2 weeks; use by specialists only; SE: pruritus, sterile abscess at injection site). Medline 2001 M11743514

Hand-Schüller-Christian syndrome (Langerhans' cell histiocytosis) Synonyms: histiocytosis X, Letterer–Siwe eosinophilic granuloma of bone. Monoclonal Langerhans-like cells are pathognomonic of this 'neoplastic', destructive, infiltrative disease in which bone, liver, skin, and spleen show lytic aggregates of eosinophils, plasma cells, and histiocytes. Lesions may show on a ⁹⁹Techetium-labelled bone scan. It occurs in children and adults, eg starting with a polyp at the external auditory meatus. Other features:

- Fever •Dyspnoea •Bone pain •Failure to thrive
- Scalp lumps •Cord compression •Skin erosions •Pustular eczema-like rash
- Proptosis •Ear discharge •Liver/spleen •Lymphadenopathy ±fits
- Stomatitis •Platelets↓ •Anaemia •Honeycomb lung

A lethal 'leukaemia' picture is seen in infants. *Treatment:* Bone surgery, steroids, cytotoxics, and radiotherapy may induce remissions. Medline 2001 M10414115 Medline 2001 M11229125

Hartnup's disease is an autosomal recessive phenotype involving a transporter for monoamino-monocarboxylic acids (gene on 11q13). Skin is thick, scaly, and hyperpigmented where exposed to light. *Signs:* Rashes, nystagmus, ataxia, bruxism, diplopia, insomnia, reflexes↑. Presentation in adults is rare. *Treatment:* Give nicotinamide and B vitamins. Medline 2003 M11394870 Medline 2001 M17955499

Hunter's syndrome (Mucopolysaccharidosis II) IDS (iduronate sulfatase) deficiency (X-linked recessive, but a new mutation in 33% so usually ♂), producing deafness, IQ↓, short stature, chronic diarrhoea, an unusual face, joint stiffness, and hepatosplenomegaly (rather like Hurler's disease, p 750 but almost always without corneal clouding, and generally pursuing a milder course). *Diagnosis* is by measuring iduronate sulfatase in the serum. *Typical age at death.* Later childhood; survival into the 30s is possible. *Pathology* Alterations at the IDS locus, mostly missense mutations. Medline 2001 M11394870

Huntington's chorea Autosomal dominant triad of progressive motor, cognitive, and emotional symptoms with loss of spiny neurones in the neostriatum—owing to excessive triplet (CAG) repeats in the huntingtin gene. Those without Huntington's have <28 repeats. **Penetrance:** A large number of repeats means the disease is more likely. 29–35 CAG repeats means no signs, but they may pass Huntington's to their children due to paternal meiotic instability. 36–39 CAG repeats means ↓penetrance. If >40 CAG repeats, 100% get Huntington's disease if they live long enough. **Early findings:** ↓Auditory & visual reaction times (needs special equipment) then mild chorea (flitting, jerky movements), odd extraocular movements, ↑reflexes, ↓rapid alternating movements. Unpredictable motor impairment is found until chorea starts. Abnormal ocular saccades may inconsistently indicate imminent manifest signs. **Late signs:** Personality change, self-neglect, apathy, clumsiness, fidgets, fleeting grimaces (may be mistaken for mannerisms), chorea, and dementia.

Ethical dilemmas surround testing, as symptoms may only start after procreation has finished. Careful pre-test counselling is vital. Medline 2001
11080230

Hunt's syndrome (Pyridoxine cerebral deficiency) Intractable neonatal fits cause death unless given pyridoxine 50 mg iv under EEG control.

Hurler's syndrome (Mucopolysaccharidosis 1H) After a few months of normal growth, there is physical & mental decline, hydrocephalus, thickened skin, hirsutism, coxa valga, and nodules over scapulae. There is deficiency of α -L-iduronidase, hence blocking degradation of dermatan sulfate and heparan sulfate, causing excess mucopolysaccharides in urine, cartilage, periosteum, tendons, valves, meninges, and cornea. **Tests:** Skull x-ray: thickened bone, absent frontal sinuses and deformed pituitary fossa. Marrow: Metachromatic Reilly bodies in lymphocytes. Death often at ≤ 10 yrs. www.txt.co.uk/full.cfm?1079

Hutchinson's triad (Congenital syphilis) Deafness+keratitis+pointed teeth

Ivemark's syndrome This is the association of congenital asplenia with ostium primum atrial septal defects (\pm pulmonary valve atresia or stenosis)

Kartagener's syndrome Primary ciliary akinesia (PCA) leads to bronchiectasis, sinusitis, σ infertility, + situs inversus (transposed heart and abdominal organs—an unexpected effect of ↓ciliary function). Otitis media and salpingitis are frequent. **Tests:** ECG: inverted P-waves in lead I, reversed R-wave progression. Mutations in dyneins (DNAI1) can cause PCA (these are large multisubunit ATPases which interact with microtubules to make cilia beat). Medline 2001
11713099

Kawasaki's disease Suspect this in anyone <9 yrs old with $T^\circ \uparrow$ for ≥ 5 days + neck lymphadenopathy (>1.5cm across), dry fissured lips, arthralgia, polymorph erythema, red eyes (uveitis), diarrhoea, strawberry tongue, or, later, finger-tip desquamation + swelling of hands/feet \pm jaundice. ►Coronary arteritis (similar to PAN) \pm infarction may occur (commoner than rheumatic fever as a cause of acquired heart disease). It may be a reaction to various infections. **Tests:** ESR/c-reactive protein \uparrow ; bilirubin \uparrow , transaminases \uparrow , α_2 -globulin \uparrow , platelets \uparrow . $\Delta\Delta$: Stevens–Johnson syndrome, measles, streps, infectious mononucleosis. **R:** γ -globulin 400mg/kg/24h ivi over 3h for 3–5 days usually brings about a rapid (diagnostic) improvement; if not, plasma exchange may help. **Follow-up:** echos; 3D coronary magnetic resonance angiography accurately defines aneurysms (non-invasive alternative when transthoracic echo quality is insufficient, and coronary angiography is too invasive). M¹11864916

Klinefelter's syndrome (xxy or xxxy polysomy + variable Leydig cell defect) A common cause of σ hypogonadism (small firm testes; small penis; 1:2000 births) may present at adolescence with psychopathy, cognition \downarrow , libido \downarrow , gynaecomastia, ↓sexual maturation. **Associations:** $T_4\downarrow$, diabetes, asthma, ?oncogenesis. Androgens and surgery for gynaecomastia may help. Life span is normal, but the arm span may exceed the body length by 10cm. M¹11864916

Klippel-Feil syndrome (Autosomal dominant or recessive; candidate KFS gene on chromosome 8) Congenital fusion of cervical vertebrae, nystagmus, deafness and CNS signs. *Mirror movements* are said to occur if *voluntary* movements in a limb are *involuntarily* mimicked by another. Progressive paraplegia may occur.

Landouzy-Dejerine (fascioscapulohumeral 4q35) muscular dystrophy

Weakness of the shoulder muscles, eg on combing the hair, appears at 12–14yrs of age. There is difficulty in closing the eyes, sucking, blowing, and whistling. Scapulae wing and the lips pout, and the facial expression is 'ironed out'. Adult myoglobin is reduced and fetal myoglobin & sarcolemma nucle are increased. Inheritance: often autosomal dominant. Medline 2001
11524495 Medline 2000
10864616

Laurence-Moon syndrome The patient is usually a boy who presents with night blindness progressing to visual loss from retinitis pigmentosa. It is distinct from the Bardet-Biedl syndrome because of the presence of paraplegia; and absence of polydactyly and obesity. Other signs: scant body hair, azoospermia, IQ↓, renal anomalies (calyceal clubbing, cysts, or diverticula; fetal lobulation; end-stage renal failure in 15%). Inheritance: autosomal recessive.

Leber's hereditary optic atrophy There is ↑risk of neoplasia + neuropathy mostly affecting males, with acute or subacute bilateral visual loss and central scotoma. A mutation (G→A) in mitochondrial DNA coding for a dehydrogenase enzyme has been proposed (a type of cytoplasmic inheritance).

Lesch-Nyhan syndrome ↓Hypoxanthine-guanine phosphoribosyl transferase (PP-ribose-P) leads to hyperuricaemia in boys (x-linked ∴ only fully expressed if ♂) with orange crystals in the nappy, then motor delay, IQ↓ (eg <65), torsion spasms, clonus, choreoathetosis, hypotonia, and fits. Compulsive, agitated, self-mutilation (foot biting, head banging, face scratching—may be unilateral) then follow gout, renal colic, and megaloblastic anaemia. Smiling aggression to others may occur. The chief defect might be ↓pain-consciousness *perhaps* related to dopaminergic denervation of D₁ receptors in cingulum bundle projections. Death is usually before 25yrs, from renal failure or infection.

Treatment: Good hydration (urine flow↑). Allopurinol prevents urate stones, but not CNS signs. It has been necessary to remove teeth to stop lip mutilation.

Lewy-body dementia Characterized by Lewy-bodies in brainstem and cortex, and a fluctuating but persisting cognitive impairment, parkinsonism, hallucinations, and visuoperceptual deficits. It is a common type of dementia. Lewy bodies are eosinophilic intracytoplasmic neuronal inclusion bodies; there is overlap between Lewy-body dementia and Alzheimer's and Parkinson's diseases, making treatment hard as antiparkinsonian agents can precipitate delusions, and antipsychotics worsen parkinsonism. Medline 2002
11841138

Li-Fraumeni syndrome This is a heritable condition in which families are highly susceptible to multiple malignancy at an early age. As well as being devastating for the families, it is of great interest to geneticists as families inherit a germ-line nonsense or oncogene-like missense mutation in one p53 allele.

Lutembacher's syndrome Secundum ASD + mitral stenosis—a rare, possibly fortuitous conjunction, unless iatrogenic, at valvuloplasty. Medline 2001
11593210

Martin-Bell (fragile X) syndrome (Semidominant; prevalence: 1:5700) This is the commonest form of inherited cognitive impairment, and is one of the commonest single-gene disorders. It is caused by a stretch of CGG-repeats within the fragile X gene, which increases in length as it is transmitted from generation to generation. Once the repeat exceeds a threshold length, no fragile X protein is produced and disease results. **Other features:** Large testes, high forehead, a big jaw, facial asymmetry, long ears, and a short temper. **Tests:** Prenatal tests are possible. Screening could be general or of high-risk groups (eg families with >1 retarded ♂). Screening for carrier status leads to labelling and stigmatization, and is rejected by some families. M10782066

Li-Fraumeni syndrome, p53, and the guardian of the genome

p53 is a tumour-suppressor gene (chromosome 17p13.1; encoding nuclear phosphoprotein, a transcription factor allowing passage through the cell cycle). In this syndrome, as only one allele is affected, development is normal until a spontaneous mutation affects the other allele. Somatic mutation of p53 occurs at both alleles in 50–80% of spontaneous human cancers. Cells with a p53 mutation do not pause in G1 (a phase in which DNA repair takes place, and faulty DNA purged), but proceed straight to S1 (DNA replication), which is why p53 protein is known as the ‘guardian of the genome’.

Note that tumours are associated with more than one syndrome, eg adrenocortical tumours are associated with familial cancer syndromes such as the Beckwith–Wiedemann and Li–Fraumeni syndromes, the Carney complex (p 647), multiple endocrine neoplasia type 1, congenital adrenal hyperplasia, and the McCune–Albright syndrome (p 754).

McCune–Albright syndrome Polyosteotic fibrous dysplasia of bone, irregular areas of skin pigmentation and facial asymmetry \pm precocious puberty.

Meckel's diverticulum Prevalence: $\leq 2\%$. ≤ 2 inches long, and >2 feet from the ileocaecal valve (antemesenteric aspect of ileum), it contains gastric and pancreatic tissue, and may be the cause of occult GI pain and bleeding (brick red stools, or dark becoming bright). This is a leading cause of rectal bleeding (\pm GI obstruction) in children. *Diagnosis*: radioisotope scan; laparotomy.

Monteggia fracture Fracture of proximal $\frac{1}{3}$ of ulna, with angulation + radial head subluxation, caused forced pronation. Open reduction/plating aids good alignment (5 weeks in plaster), so as not to impair pronation. Wait for full union (~ 12 weeks) before normal arm use.

Medline 2002
M10695863

Morquio's syndrome (Mucopolysaccharidosis IV) Defective degradation of keratan sulfate (it is excreted in the urine) causes stature \downarrow ; deafness; weakness; a coarse, broad mouth; widely spaced teeth; pectus carinatum; aortic regurgitation—but not mental retardation. Autosomal recessive.

Moyamoya disease Vascular disease at the Circle of Willis causes strokes or TIAs (alternating hemiplegia, dyspraxia, headache). The typical patient is a Japanese girl, with triggering infection (eg tonsillitis) or hyperventilation ($\text{CO}_2\downarrow$ causes vasoconstriction). Digital subtraction angiography shows collateral vessel formation 'like a puff of smoke'. MRI/CT shows multiple infarctions in watershed areas. Bypass surgery may be possible.

Medline 2002
M10695863

Niemann–Pick disease After months or years of normal growth there is physical and mental decline, wasting \pm hepatosplenomegaly from abnormal metabolism of sphingomyelin. Other signs: brown skin patches \pm a cherry red spot on the macula. There are 5 types; eg, type D is caused by a G3097 \rightarrow T transversion in NPC1 gene.

Medline 2002
M9634529

Noonan syndrome Hypertrophic cardiomyopathy, VSD/ASDs, bruising (APTT \uparrow and \downarrow factors VIII, XI, XII), ptosis, down-slanting eyes, low-set ears, webbed neck. Height, IQ, and hearing are below average, but not severely so. *Inheritance*: Autosomal dominant (gene region 12q24). *Prevalence*: 1:5000.

Othello syndrome (Morbid jealousy) A lover believes against all reasons that his beloved is being sexually unfaithful, and may also think that she is plotting against him, deliberately making him impotent. He may engage a detective to spy on her, and examine her underwear for signs of sexual activity. It may be primary or secondary (to alcohol, schizophrenia, neurosis, or depression).

► Get psychiatric help: it may be deadly '... sometimes even without my having seen Albertine again ... there would flash from my memory some vision of her with Gisèle in an attitude which had seemed innocent to me at the time; it was enough now to destroy the peace of mind I had managed to recover, I had no longer any need to go and breath dangerous germs outside, I had, as Cottard would have said, supplied my own toxin'. Proust, *Sodome & Gomorrhe*

Patau's syndrome (Trisomy 13) The head and eyes are small: \pm defects such as absent corpus callosum and a single ventricle. Heart lesions, polycystic kidneys and cleft lip/palate are common. Hands show flexion contractures \pm polydactyly with narrow fingernails. *Prevalence*: 1/7500 births.

Pick's dementia This often presents later than Alzheimer's disease—eg with character change associated with frontal lobe signs such as gross tactlessness. Lack of restraint may lead to stealing, practical jokes, and unusual sexual adventures. Social graces are replaced by fatuous euphoria and jargon dysphasia, before intellectual and CNS deterioration occurs.

Pierre Robin syndrome Neonatal difficulty in feeding and breathing due to micrognathia (short chin) \pm cleft palate or eye abnormality. Prevent the tongue slipping back by nursing on the belly (chest elevated on a pillow).

Hyperendocrinopathies, McCune–Albright syndrome, and the Taiwanese giant

In the McCune–Albright syndrome, precocious puberty is not the only endocrinopathy: hyperthyroidism and Cushing's have also been reported. In the case of the 'Taiwanese giant' (an unfortunate name) excess growth hormone production has also been found. M 1998 9678473 Deformities, fractures, and pain further complicate the picture (sometimes ameliorated by iv pamidronate). The craniofacial fibrous dysplasia may encroach on the optic nerve, causing visual problems. GnRH analogues have been used (experimentally) to treat the precocious puberty. Medicine 1998 M 9642633

The cause is thought to be a mutation of the *GNAS 1* gene coding the α subunit of the stimulatory guanine-nucleotide binding protein, G-protein, which activates adenylate cyclase ($\therefore \uparrow$ intracellular cyclic AMP). M 1998 9578646

Pompe's glycogen storage disease (type II; GSDII) \uparrow Lysosomal $\alpha_{1,4}$ -glucosidase activity leads to weakness and failure to thrive in early (or later) life. IQ \downarrow and chest infections occur. Glycogen accumulates in heart; muscle; CNS; liver; kidney; adrenals. Few survive beyond infancy. Mutations include: a G residue insertion in exon 16 (InsG2242), a deletion of 20 nucleotides in exon 4 delta, and a nonsense mutation in exon 16 (G2237A-Trp746Stop). M¹¹ 1020668

Prader-Willi syndrome (pws) *Cause:* Loss of the paternal contribution of the proximal part (15q11–q13) of the long arm of chromosome 15 (deletion, disomy, and imprinting mutations). *Prevalence:* 1:25,000. *Signs:* Hypotonia, epilepsy, failure to thrive, and sleepiness. The child usually has blue eyes and blond hair. Later, hyperphagia (eg eating dog biscuits, or shop-lifting food), obesity, hypogonadism, short stature, hyperglycaemia, cor pulmonale, and sleep apnoea occur. Toddlers described as passive, autistic, or introverted may go on to show unstable mood disorder. Those who are extrovert may develop psychosis. *Tests:* EEG: slow spike & wave activity. M¹¹ 1185185

Ramsay Hunt syndrome (*Herpes zoster oticus*) Severe otalgia, usually in the elderly, precedes facial nerve palsy. Zoster vesicles appear around the ear, in the deep meatus (\pm soft palate & tonsillar fossa). There may be vertigo and sensorineural deafness which are both slow to resolve. M¹¹ 1180037

Rett disorder A neurodevelopmental disorder of repetitive hand-wringing with loss of purposeful hand use, eg at 2–3yrs old, with mental and motor regression \pm partial seizures \pm autism (p 392) \pm arrhythmias and apneustic or Valsalva breathing—from an X-linked mutation affecting girls: it is lethal to the σ^* fetus). MECP2 gene mutations are the cause in \sim 70%. M¹¹ 11313764 M¹¹ 11420195

Diagnostic using DSM IV is complex. www.geocities.com/HotSprings/9647/dsm-crit.htm#retts. It is similar to cretinism and fragile X (p 752) as there are defects in activity-dependent neuronal plasticity (how neuronal activity shapes new circuits) causing faulty intracellular signaling pathways with failure of linking of cell surfaces with nuclear events for gene expression (signaling pathways so disrupted are involved in learning, memory, and behaviour as well as in 'synaptic proliferation and pruning' occurring during normal development). M¹¹ 11407158

Reye's syndrome *Presentation:* Vomiting, fever, hypotonia, stupor/coma, liver failure. It may have an infectious cause and/or be related to aspirin, antiemetic or antihistamine use by children. Median age of onset: ^{uk} 14 months. *Tests:* Glucose \downarrow ; transaminases \uparrow ; blood ammonium \uparrow (levels correlate with survival); INR \uparrow . Liver biopsy: swollen, pleomorphic mitochondria (ATP \downarrow gluconeogenesis & ureagenesis \downarrow). $\Delta\Delta$: Inherited metabolic disorders (IMD).

Russell Silver syndrome Asymmetrical growth/hemihypertrophy with small stature and precocious puberty. Association: Wilms'. Aberrant genomic imprinting may be to blame. www.asri.edu/gene-tics/brochure/agh/news/oct96/imprinting.htm

Shakhonovich's syndrome (*Hypokalaemic periodic paralysis*) Attacks precipitated by: stress, menstruation, cold, large carbohydrate meals, rest after exercise or liquorice—typically starting in early morning, with flaccid paralysis, beginning in the legs, and spreading upwards. Eye movements, swallowing, and speech are unaffected. Recovery takes place over 1–24h. Typical age 7–21yrs; may have a history of migraine. During attacks muscles feel firmer than usual. Tendon reflexes are diminished. *Tests:* Serum K^+ and phosphorus usually fall during attacks; urate may rise. Glycosuria may be present. $wcc \uparrow$ + eosinopenia. Autosomal dominant, with implication of a gene coding $\alpha 1$ -subunits of skeletal muscle L-type calcium channel. Sporadic cases also occur. *Treatment:* Spironolactone + very low Na^+ diet may prevent attacks. M¹¹ 11808345

Staging and treatment of Reye's syndrome

The point of staging is to recognize stage 2 (or worse) which should prompt rapid referral to a tertiary centre with facilities for monitoring icp (eg with subarachnoid bolts) and intra-arterial pressure. Stage 2 criteria: inappropriate verbalizing, combative or stuporose, normal posture, purposeful or non-purposeful response to pain, sluggish pupillary responses, intact eye reflex.

Management: ► Correct hypoglycaemia; set up a *continuous* ivl of 10–15% glucose. Fluid restriction; do blood glucose every 2h; give vitamin k 0.25mg/kg slowly iv (monitor prothrombin time); lower icp—see p 256. Aim for 40mmHg cerebral perfusion pressure (= systolic BP minus icp). Control fever and seizures.

Mortality: ~31%. Since abandoning aspirin in children, incidence has fallen to ≤ 1 –6 patients/yr/ 10^6 children <16yrs.

www.nejm.org/content/1999/0340/0018/1377.asp

Still's disease Typically this juvenile chronic arthritis subtype (JCA; OTM-3 2963) presents with systemic upset in a prepubertal girl with: mono- or polyarticular synovitis (eg with progressive erosion of cartilage) \pm fevers, pericarditis, iridocyclitis, pneumonitis (lung biopsy specific), lymphadenopathy, splenomegaly. Rheumatoid factor is not present in the blood. Medline 1992 11807193

Other subgroups are juvenile ankylosing spondylitis, psoriatic arthritis and ulcerative colitis-associated arthritis, and juvenile-onset rheumatoid arthritis (here Rh factor +ve, and less likely to present as systemic upset).

Adult-onset Still's disease (AOSD): diagnostic criteria—all of: daily fever $>39^{\circ}\text{C}$; arthralgia/arthritis; Rh factor and antinuclear factor –ve plus any 2 of: wcc $>15 \times 10^9/\text{L}$; rash; serositis (pleural or pericardial); hepatomegaly; splenomegaly; lymphadenopathy—provided that endocarditis, leukaemia and sarcoidosis are excluded. As with all rheumatology, classification is muddled, and unexpected complications may occur (DIC, liver failure; renal failure; amyloid; cardiac tamponade; sterile endocarditis; peritonitis).

Tests: WCC \uparrow ; ESR \uparrow ; mild anaemia; ferritin \uparrow ; LFTs \uparrow ; albumin \downarrow ; echo. Medline 1995 8746917

Treatment: Mild exercise is followed by one hour's rest per day. If the hips are affected, physiotherapy aims to prevent contractures by encouraging extension (eg lying prone on the floor to watch tv). Splinting, traction, and non-weight-bearing exercises may be used. Hot baths relieve morning stiffness. Give aspirin, up to 80mg/kg/day po pc to give levels $<250\text{mg/L}$ (beware of liver and CNS toxicity). Alternative: Naproxen 5mg/kg/12h po pc. If there is severe systemic disturbance, prednisolone 0.5mg/kg/day po may be needed. For those with unremitting destructive disease, penicillamine, gold, hydroxychloroquine, and infliximab (in those with AOSD) may be tried. Surgery may be needed to conserve joint function. Medline 2002 11890655

Sydenham's chorea (St Vitus' dance) This may start with emotional lability and a preference for being alone (\pm attention span \downarrow). Next comes purposeless movement, exaggerated by tension, and disappearing on sleep, with clumsiness, grimacing, a darting lizard's tongue and unclear speech (OHCM p 52). It is part of rheumatic fever (in $<5\%$, p 194), and may be the only feature, appearing up to 6 months after clinical and lab signs of streptococcal infection have abated. The term **PANDAS** (pediatric autoimmune neuropsychiatric disorder associated with streptococcus) aims to describe a putative subset of obsessive-compulsive disorder and Tourette's syndrome (OHCM p 726) that bears some resemblance to Sydenham's chorea. Medline 2002 10807957

$\Delta\Delta$: Wilson's disease, juvenile Huntington's, thyrotoxicosis, SLE, polycythaemia, $\text{Na}^+\downarrow$, hypoparathyroidism, kernicterus, encephalitis lethargica, subdural haematoma, alcohol, phenytoin, neuroleptics, hereditary chorea, neuroacanthosis. **R_x**: (if desired) carbamazepine; sodium valproate. Medline 2002 11890655

Syme's amputation An amputation immediately proximal to the ankle.

Tay-Sachs disease This is an autosomal recessive gangliosidosis (Type I) which affects $\sim 1:4000$ Ashkenazic Jewish births. It is a disease of grey matter. There is reduced lysosomal hexosaminidase A. Low levels of enzyme are detectable in carriers. Children are normal until ~ 6 months old, when developmental delay, photophobia, myoclonic fits, hyperacusis and irritability occur. Ophthalmoscopy: cherry-red spot at macula. Death usually occurs at 3–5yrs of age. Prenatal diagnosis may be made by amniocentesis.

Tolosa-Hunt syndrome A lesion in the superior orbital fissure produces unilateral ophthalmoplegia with disordered sensation (1st branch of V).

Treacher-Collins' syndrome Lower lid notching, oblique palpebral fissures, flattening of malar bones \pm hypoplastic zygoma. If these are associated with mandibular defects, ear defects, and deafness, it is called Franceschetti's syndrome. Mutations in the TCOF1 gene may be to blame.

Turner's syndrome (xo karyotype or mosaic, eg 45X/46,XX or 45,X/47,XXX.)

Prevalence: 1:2500 girls. **The Patient:** Girls typically lack a sex chromosome, and this is associated with short stature (<130cm) \pm , hyperconvex nails, wide carrying angle (cubitus valgus), inverted nipples, broad chest, ptosis, nystagmus, webbed neck, coarctation of the aorta, left heart defects, leg lymphoedema. Gonads are rudimentary/absent and puberty may not occur.

Typical mode of death: Heart disease. **Rx:** Somatropin (human GH; 0.6–2u/kg/week sc, or in divided nightly doses) adds a median of 5.1cm to final stature (6.4cm if GH + oxandrolone \leq 0.1mg/kg/day); don't give if epiphyses are fused. **NB:** GH may prolong the physiological state of insulin resistance observed in normal puberty (resultant hyperinsulinaemia may \uparrow anabolic effects of insulin on protein metabolism during puberty). ^{Medline 2001} M11529402 If hypogonadic, HRT, eg 2mg estradiol valerate + levonorgestril 75 μ g at night, can start at 13yrs. \blacklozenge

Associations: Crohn's disease; chronic liver disease. ^{Medline 2001} M11572325 ^{Medline 2001} M11531930

Von Gierke's syndrome (type Ia glycogen storage disease; GSD 1a) is the most common and severe of the glycogenoses. **Inheritance:** Autosomal recessive (G6Pase gene mutation, causing \downarrow glucose-6-phosphatase). **Signs:** Hepatomegaly, kidney enlargement, growth retardation, hypoglycaemia, lactic acidaemia, hyperuricaemia, failure to thrive, lumbar lordosis, hyperlipidaemia, adiposity, xanthomata over joints and buttocks, and a tendency to bleed. **Complication:** Hepatic adenoma; hepatocellular carcinoma. ^{Medline 2002} M11477366

Werner's syndrome (ws; progeria) is characterized by precocious ageing after puberty, with diabetes, cataracts and scleroderma-like skin changes. The complex molecular and cellular phenotypes of ws involve features of genomic instability and accelerated replicative senescence. The gene involved (WRN) was recently cloned, and its gene product (WRNp) is a helicase. Helicases play important roles in a variety of DNA transactions, including DNA replication, transcription, repair, and recombination, and in ws unwinding of DNA pairs is disordered. ^{Medline 1999} M10436020 ^{Medline 2001} M11380620

Winkler's disease (chondrodermatitis nodularis chronica helici) The typical patient is a middle-aged man with a chronic, scaly, ulcerated nodule on the helix of the right ear (or the antehelix in women)—hence its synonym: chondrodermatitis nodularis heliciis chronicis. Multiple lesions may occur along the ear's rim. **Cause:** Recurrent minor trauma is thought to be important. It occurs more commonly on the ear on the side of sleeping. Telephone operators are at risk, as are nuns wearing tight head dresses. It is benign. **Treatment:** Surgical excision, or use of the argon laser. ^{Medline 1999} M10500520

Wiskott-Aldrich syndrome is a severe X-linked primary immunodeficiency (p 201) with thrombocytopenia, small platelets, eczema, recurrent infections, autoimmune disorders, IgA nephropathy, and risk of hematopoietic malignancies. Without marrow transplant, most will die before adulthood. DNA linkage helps with prenatal diagnosis. The responsible gene has been found, along with its product, known as Wiskott-Aldrich syndrome protein. Prenatal tests include direct gene analysis with single strand conformation polymorphism (sscp) and heteroduplex formation (HD). ^{Medline 2001} M11892069

11 Anaesthesia

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Other relevant pages: Pain relief in labour (p 114); resuscitation after delivery (p 230); neonatal ITU (p 232); ventilating neonates (p 234); major injury (p 684); choosing the correct dose of lidocaine (=lignocaine) according to body weight (p 687); pain relief in children (p 678); Bier's block (p 700).

Introduction

Anaesthesia has evolved from humble origins. It was 1842 when CW Long gave ether, but he failed to report this landmark in the relief of pain. Then, in 1844, Horace Wells used nitrous oxide for tooth extraction, and in 1846 WTG Morton gave the first surgical anaesthetic with ether. It is now a highly specialized and sophisticated specialty in its own right. While the triad of anaesthesia (hypnosis, analgesia, and muscle relaxation) remains the fundamental principle behind general anaesthesia for surgery, the rôle of the anaesthetist has expanded to encompass not only the provision of ideal operating conditions for surgery, but also intensive care, resuscitation, alleviation of acute and chronic pain, and anaesthesia for diagnostic procedures. A detailed knowledge of general medicine, physiology, pharmacology and the physical properties of gases and the workings of the vast array of anaesthetic equipment are essential, in order to practise well.

►We emphasize that this short chapter is no substitute for a specialist text or for experience on the ward and in theatre. The main aim is to enable understanding of the issues anaesthetists face, and to prepare one's mind for intelligent discussions on anaesthetic issues.

Pre-operative care

Aims Principally to ensure that the patient gets the right surgery. Have the symptoms and signs changed? If so, inform the surgeon. Also:

- 1 To assess and balance the risks of anaesthesia.
- 2 To ensure that the patient is as fit as possible.
- 3 To decide on the type of anaesthesia and analgesia.
- 4 To allay all anxiety and pain.

The pre-operative visit Assess cardiovascular and respiratory systems, exercise tolerance, existing illnesses, drug therapy, and allergies. Assess past history—of myocardial infarction, diabetes, asthma, hypertension, rheumatic fever, epilepsy, jaundice. Assess any specific risks—eg is the patient pregnant? Any dental problems, or history of GI reflux? Have there been any anaesthetic problems (nausea, DVT)? Has he had a recent GA?

Family history Ask about malignant hyperpyrexia (p 772); dystrophia myotonica (OHCM p 386); porphyria; cholinesterase problems; sickle-cell disease (test if needed). Does the patient have any specific worries?

Drugs Ask about allergy to any drug, antiseptic, plaster, and latex.

ACE inhibitors: No special action required.

Anticoagulants: Know the indication. Do INR; switch warfarin to heparin 24h pre-op. Avoid epidural/spinal blocks. Beware regional anaesthesia.

Anticonvulsants: Give the usual dose up to 1h before surgery. Give drugs iv (or by NGT) post-op, until able to take oral drugs. Sodium valproate: an iv form is available (give the patient's usual dose). Phenytoin: give iv slowly (<50mg/min). IM phenytoin absorption is unreliable.

Antibiotics: Neomycin, aminoglycosides, colistin & tetracycline prolong neuromuscular blockade, even depolarizing neuro-muscular blockers.

Beta-blockers: Continue up to and including the day of surgery as this precludes a labile cardiovascular response.

Contraceptive Pill: See BNF Stop 4 weeks before major (or leg) surgery.

Digoxin: Continue up to and including morning of surgery. Check for toxicity and do plasma K^+ . Suxamethonium \uparrow serum K^+ by $\sim 1\text{mmol/L}$, and can lead to ventricular arrhythmias in the fully digitalized.

Diuretics: Beware hypokalaemia. Do u&E (and bicarbonate).

HRT: Consider stopping before orthopaedic surgery. Use thromboprophylaxis (heparin and stockings).

Insulin: If on long acting preparations stabilize pre-operatively on short-acting. iv 5% dextrose and insulin or an insulin infusion on the morning of theatre allows accurate control. See OHCM p 458.

Levodopa: Possible arrhythmias when the patient is under GA.

Lithium: Get expert help; it may potentiate neuromuscular blockade and cause arrhythmias. Beware post-op toxicity \pm u&E imbalance: p 354.

Monoamine oxidase inhibitors: Get expert help as interaction with narcotics and anaesthetics may lead to hypotensive/hypertensive crisis.

Ophthalmic drugs: Anticholinesterases used to treat glaucoma (eg ecothiopate iodine) may cause sensitivity to, and prolong duration of, drugs metabolized by cholinesterases, eg suxamethonium.

Steroids: If the patient is on or has recently taken steroids give extra cover for the peri-operative period (p 763).

Tricyclics: These enhance epinephrine (=adrenaline), exerting anticholinergic effects causing tachycardia, arrhythmias and low BP.

Pre-operative examination and tests

It is the anaesthetist's duty to assess suitability for anaesthesia. Be alert to chronic lung disease, BP↑, arrhythmias, and murmurs (endocarditis prophylaxis needed?—p 194). In rheumatoid arthritis do a lateral cervical spine x-ray to warn about difficult intubations. Post-op chest infections are *six times* more likely in smokers. Stopping smoking even 1 day pre-op is of benefit.

The *ward doctor* assists with a good history and examination—and should also reassure, inform, and obtain written consent from the patient (eg get consent for orchidectomy in orchidopexy procedures, inform thyroidectomy patients of nerve damage risk, and retrograde ejaculation with prostate surgery). Other concerns for the house officer to address include:

- DVT prophylaxis (p 654)
- Bowel preparation
- On-table x-rays
- Frozen section (tell pathology)
- Skin preparation
- Post-op physio

Consent UK law implies we are either fully competent or 'incompetent' but things are not so clear-cut, so... **1** Use only words the patient understands. **2** Ensure he believes your facts and can retain the pros and cons long enough to inform his decision. **3** Make sure his choice is free from pressure from others. A patient may complain if: • He is unaware of what will happen • He has not been offered *all* the options • He was under the influence of drugs at the time of consent • He changed his mind • He was not told the treatment was experimental • A second opinion has been denied • Details of his prognosis were glossed over.

Remember: the patient is not on a conveyor belt. If in doubt, get legal help.

Tests Be guided by the history and examination.

- In most patients, do FBC, ward test for blood glucose, and U&E—especially important if the patient is: —On diuretics —A diabetic —A burns victim —Has hepatic or renal disease —Starved —In ileus —Parenterally fed
- If Hb < 10g/dL tell anaesthetist. Investigate/treat as appropriate.
- Group & save for all major surgery; crossmatch according to local Blood Transfusion Service guides, depending on pre-op Hb and type of surgery.
- Tests: LFT in jaundice, malignancy, or alcohol abuse. Amylase if needed. Blood glucose in diabetic patients (*OHCM* p 458). Drug levels as appropriate (eg digoxin). Clotting studies in liver disease, DIC, massive blood loss, already on warfarin or heparin. Contact lab as special bottles are needed. HIV, HBsAg in high-risk patients—after appropriate counselling. Sickie test in those from Africa, West Indies, or Mediterranean area—and others whose origins are in malarial areas (including most of India). Thyroid function tests in those with thyroid disease.
- CXR: if known cardiorespiratory disease, pathology or symptoms.
- ECG: those with poor exercise tolerance, angina etc., BP↑, past rheumatic fever/Kawasaki's (p 750), age >50 (high incidence of 'silent' ischaemia).

Preparation ▶ Fast the patient: ie no water for 2h pre-op, no milky drinks/squash for 4h, and no solids for 6h.

Use nasogastric tube as indicated: insert *before* induction to avoid risk of aspiration at induction. (Technique: *OHCM* p 742.) Catheterize as indicated.

ASA classification (American Society of Anesthesiologists)

- 1 Normally healthy.
- 2 Mild systemic disease, but with no limitation of activity.
- 3 Severe systemic disease that limits activity; not incapacitating.
- 4 Incapacitating systemic disease which poses a threat to life.
- 5 Moribund. Not expected to survive 24h even with operation.

You will see space for an ASA number on standard anaesthetic charts. It is an index of health at the time of surgery; the prefix **E** denotes emergencies.

Premedication

The patient should be aware of what will happen, where he will waken and how he will feel. Premedication aims to allay anxiety and contribute to a smooth induction of anaesthesia by decreasing secretions (much less important than when ether is used), promoting amnesia, analgesia, and decreasing vagal reflexes. *Timing:* 2h pre-op for oral drugs; 1h pre-op if IM.

Examples for the 70kg man

- Lorazepam 2–3mg po.
- Temazepam 10–40mg po.
- Diazepam 5mg po.

Some anaesthetists still use the traditional IM premeds. Examples:

- Morphine 10mg IM and atropine 0.6mg IM.

Examples for children

- Alimemazine = trimeprazine (child 2–7y) 2mg/kg as a syrup.
- Always use oral premeds in children as first choice.
- EMLA® local anaesthetic cream on at least 2 sites that may be used for IV access, and at least 1h prior to venepuncture (early vasoconstriction makes venepuncture difficult before this).

Specific premedication

- Antibiotics
- Bronchodilators, eg salbutamol nebulizer
- Nitrate patches, eg Transiderm Nitro®—esp. useful in cardiac surgery.
- Steroids, eg minor operations 100mg of IM hydrocortisone 1h pre-op and 6h post-op. Major operations hydrocortisone 100mg/6h IM. Consider if adrenal insufficiency or adrenal surgery; steroid therapy for more than 2 weeks prior to surgery; or steroid therapy for more than 1 month in the year preceding surgery.
- Antacids.
- Drugs to prevent gastro-oesophageal reflux, eg sodium citrate, metoclopramide, or ranitidine started the evening before surgery.

Common reasons for cancellation

- Cold or recent viral illness if still symptomatic.
- Recent myocardial infarction (significant ↑ in mortality if GA is given within 6 months—believed to be related to degree of cardiac failure).
- Patient not adequately fasted.
- U&E imbalance (particularly K⁺); anaemia.
- Inadequate preparation (results not available, not crossmatched).
- Patient not in optimum condition—eg poor control of drug therapy (digoxin, thyroxine, phenytoin); exacerbation of illness.
- Undiagnosed or untreated hypertension.
- Inadequate theatre time.

Equipment

Careful checking of equipment is vital before any anaesthetic or sedative procedure. The essentials are:

- Tilting bed or trolley (in case of vomiting).
- High-volume suction with rigid Yankaeur, and long suction catheters.
- Reliable oxygen supply, capable of delivering 15L/min.
- Self-inflating bag with oxygen reservoir, non-rebreathing valve, and compatible mask (a 'bag-valve-mask' system).
- Oropharyngeal, nasopharyngeal, and laryngeal mask airways.
- A range of anatomical face mask sizes with harness.
- Endotracheal tubes (range) and catheter mount.
- Anaesthetic circuit.
- Laryngoscope with range of blade sizes, spare bulbs, and batteries.
- Intravenous infusion cannulae and fluids.
- Anaesthetic and resuscitation drugs and anaesthetic gases.
- Defibrillator.
- Monitoring equipment (may include pulse oximeter and end tidal CO₂ monitor, p 772).

Inhalational agents

These are the vapours which in clinically useful concentrations help to maintain anaesthesia and decrease awareness. In Britain they are generally added to the fresh gas flow by passing a fraction of the carrier gas (generally N₂O/O₂) through a plenum vaporizer.

Halothane This gas has little analgesic effect. It decreases cardiac output (vagal tone ↑, leading to bradycardia, vasodilation, and hypotension). It sensitizes the myocardium to catecholamines (beware in patients with arrhythmias; surgical infiltration with local anaesthetic and epinephrine). Halothane has been replaced by safer inhalational agents, partly because of problems with jaundice and repeated use.

Isoflurane This is a halogenated ether. Theoretically induction should be quick, but isoflurane is irritant, so coughing, laryngospasm, or breath-holding may complicate the onset of anaesthesia.

Sevoflurane This is a halogenated ether which is well-tolerated. It is the agent of choice for inhalation induction of general anaesthesia with low blood : gas solubility.

Desflurane is another halogenated ether with a pungent smell, rapid onset of anaesthesia, and quick recovery.

Stopping inhalation reverses all the above effects—except for hepatitis resulting from drug metabolism.

The ideal iv anaesthetic agent

The ideal iv agent would be stable in solution, be water-soluble and have a long shelf-life. It would be painless when given iv; non-irritant if injected extravascularly (with a low incidence of thrombosis) with some pain (as a warning) if given intra-arterially.

- It should act rapidly within one arm–brain circulation.
- Recovery should be quick and complete with no hangover effect.
- It should provoke no excitatory phenomena.
- Analgesic properties are advantageous.
- Respiratory and cardiovascular effects should be minimal.
- It should not interact with other anaesthetic agents.
- Hypersensitivity reactions are not ideal.
- There should be no post-op phenomena, eg nausea or hallucinations.

This perfect drug does not exist.

IV anaesthetic agents: thiopental sodium

Thiopental (=thiopentone) sodium ($t_{1/2} \approx 11\text{h}$) When mixed with water to give a 2.5% solution this barbiturate is stable for 24–48h, so make it up daily. It has a rapid onset of action (arm–brain circulation time about 30sec). Effects last 3–8min. Awakening is due largely to redistribution, not metabolism. Some 30% of the injected dose is still present in the body after 24h, giving rise to the hangover effect. Patients must not drive, operate machinery etc. within 24h. One mL of 2.5% solution contains 25mg/mL.

Dose: eg 4–6mL of 2.5% solution (3–5mg/kg, but less in the elderly and the premedicated, and more in children who generally require 6–7mg/kg, but some children need much less, eg 2mg/kg.) Subsequent doses are cumulative. Note: thiopental (=thiopentone) sodium interacts with other drugs eg increase dose in those who consume much alcohol. Uses: induction of GA; it is also a potent anticonvulsant.

Contraindications: ● Airway obstruction ● Barbiturate allergy ● Fixed cardiac output states ● Hypovolaemia/shock ● Porphyria.

► Problems: Intra-arterial injection produces pain and blanching of the hand/limb below the level of injection due to arterial spasm, followed by ischaemic damage and gangrene—frequently following inadvertent brachial artery puncture in the antecubital fossa.

Treatment: 1 Leave the needle in the artery and inject a vasodilator such as papaverine. 2 Perform, or ask an experienced colleague to perform, brachial plexus or stellate ganglion block. (These measures should dilate vessels and reduce ischaemia.) 3 Heparin iv to stop thrombus forming. 4 Give pain relief. 5 Postpone surgery unless desperate.

Extravascular injection produces severe pain and local tissue necrosis. Infiltrate with hyaluronidase 1500iu dissolved in 1mL water for injection through the cannula.

Other iv anaesthetic agents (See p 772)

Etomidate ($t_{1/2} = 3.5\text{h}$) This is a carboxylated imidazole. It is used as an induction agent. Dose: 0.1–0.3mg/kg. Histamine release is not a feature; rapidity of recovery and cardiovascular stability are; therefore it is suitable for day-case surgery, the elderly, and those with compromised cardiovascular systems. Be aware that this may induce involuntary muscle movements, nausea, and adrenal suppression.

Ketamine (used mainly for paediatric anaesthesia) Dose: 0.5–2mg/kg; $t_{1/2} = 22\text{h}$. This is a phencyclidine derivative. There may be delay before onset of sleep. Hypertonus and salivation are problems, but there is some maintenance of laryngeal reflexes (do not rely on this). Recovery is slow. Emergence phenomena are troublesome (delirium, hallucinations, nightmares; all made worse if the patient is disturbed during recovery). Cardiac output is unchanged or increased. It is a good 'on site' or 'in the field' agent, as it can be given IM and produces profound analgesia without compounding shock. ▶ Avoid in the hypertensive patient, those with a history of stroke, or raised intracranial pressure (further ↑ produced), patients with a recent penetrating eye injury (risk of ↑ intraocular pressure), and psychiatric patients. Avoid epinephrine infiltrations.

Propofol This phenol derivative is the most widely used iv anaesthetic agent and is formulated in soya oil and egg phosphatid.

• **Dose example:** 2mg/kg iv at 2–4mg/sec. It is not good for day surgery (delayed convulsions). ▶ Rapid injection can cause cardiovascular depression (↓BP), and respiratory depression can occur when combined with iv narcotics. For sedation: 1.5–4.5mg/kg/h iv. It acts and is metabolized quickly. Wake-up is caused by redistribution, not metabolism. It is used in induction and maintenance of GA, and for sedation during regional anaesthesia, short procedures, and as a sedative in ITU. ci: the extremities of age; egg allergy. NB: dose depends partly on premed use: with midazolam premeds, propofol requirements are less by up to 20%. N=24
2001 medline-11394510

- Its favourable recovery characteristics and antiemetic effects have made it widely popular.
- Once opened, use ampoules or discard, because of the risk of bacterial growth. Propofol in rubber-capped vials has antibacterial properties.
- Pain on injection occurs in up to 40% of patients. This can be minimized by either adding a small amount of lidocaine (=lignocaine) (2mg of 1%) to the propofol, or by pre-injection of a similar volume of local anaesthetic.

Neuromuscular blockers

► Never give a muscle relaxant in the presence of a compromised airway.

These drugs act on the post-synaptic receptors at the NMJ (neuromuscular junction). There are two main groups:

1 Depolarizing agents eg suxamethonium (=succinylcholine, Scoline®). These drugs depolarize the post-synaptic membrane, causing paralysis by inhibiting the normal membrane polarity. They are partial agonists for acetylcholine and cause initial fasciculation, liberation K^+ , enough to raise the plasma K^+ by $\sim 0.5\text{--}1.0\text{mmol/L}$ (beware in paraplegia and burns!), myoglobin and creatine kinase. Beware that K^+ liberation is increased with multiple sclerosis, Guillain-Barré, stroke, and crush injury. They also increase intra-ocular pressure (eg increase risk of vitreous extrusion). 30% of patients get post-operative muscle pains. Suxamethonium is an ideal intubating agent: it has a rapid onset, a short duration of action (2–6min), and produces good relaxation. Note: a second dose, if required, should be preceded by atropine, as the vagotonic effects of suxamethonium can lead to profound bradycardia. Beware suxamethonium (Scoline®) apnoea (p 772). Dose of suxamethonium: eg $0.5\text{--}1\text{mg/kg}$ iv. Bradycardia related to the 1st dose is a problem in children.

2 Non-depolarizing agents These drugs compete with acetylcholine at the NMJ—but without producing initial stimulation (see suxamethonium above). Repeated doses may be given without atropine. Their action can be reversed by anticholinesterases (neostigmine). They are used during balanced anaesthesia to facilitate IPPV and surgery. Length of action and side-effects will govern anaesthetists' choices. Examples of the more common agents are:

Vecuronium Lasting 20–30min, it is used if cardiovascular stability is important. No ganglion-blocking effect. Dose: $80\text{--}100\mu\text{g/kg}$ iv then $20\text{--}30\mu\text{g/kg}$ iv as needed. Starting dose for infants ≤ 4 months: $10\text{--}20\mu\text{g/kg}$. If over 5 months, as adult dose, but high intubation dose may not be needed for the age group between 5 months and 1 year. Known as the 'cleanest' of these three agents due to least histamine release.

Atracurium Lasting $\sim 20\text{min}$ this causes histamine release so avoid in asthma. Metabolism is by Hoffman Elimination (spontaneous molecular breakdown), so it is the drug of choice in renal and liver failure. Dose: $300\text{--}600\mu\text{g/kg}$ iv then $100\text{--}200\mu\text{g/kg}$ iv as needed.

Pancuronium Long acting ($\sim 1\text{h}$), vagal blockade and sympathomimetic action. Dose: $50\text{--}100\mu\text{g/kg}$ iv then $10\text{--}20\mu\text{g/kg}$ iv as needed. (No longer used in some anaesthetic practice.)

Other non-depolarizing agents in use include cisatracurium, rocuronium, and mivacurium.

Note on neuromuscular blockers in those with myasthenia In general, these patients are resistant to suxamethonium and sensitive to non-depolarizing relaxants (action may be prolonged: lower doses may be needed). Warn patients that mechanical ventilation may be needed post-op. Liaise closely with a senior anaesthetist.

Practical conduct of anaesthesia: I

The practitioner administering the anaesthetic is responsible for the suitability of the surroundings, the adequacy of the available equipment, and his own competence to deal with potential complications. Equipment *must* be checked before even the shortest anaesthetic procedure.

Induction May be gaseous or iv (im possible with ketamine).

Gaseous:

- Either start with nitrous oxide:oxygen 60%:40% mixture. In children, it is less frightening to start with a hand cupped from the end of the circuit onto the face than to apply the mask direct to face.
Or give sevoflurane in oxygen according to patient's age and clinical state for rapid induction of general anaesthesia.
- Monitor vital signs closely, as the time factor separating a struggling child from a flaccid, apnoeic, overdosed child is short.
- Establish iv access when and where appropriate, as soon as asleep.

Indications for gaseous induction:

- Any patient with airway obstruction (actual or potential, eg foreign body, tumour, or abscess).
- At the patient's request.
- Difficult iv access.

Intravenous:

- Establish iv access.
- A sleep-inducing dose of, eg thiopental (=thiopentone) sodium, is injected after a 2mL test dose to detect inadvertent arterial cannulation (p 764).
- Beware! Stimulation before anaesthesia is established can have drastic consequences (coughing, breath-holding, laryngospasm). Remember, noise is a stimulus too.

Airway control This is maintained either by holding a mask onto the face, by inserting a laryngeal mask airway, or by intubation (p 771). To prevent airway obstruction, standard chin lift and/or jaw thrust manoeuvres are used. It may be facilitated by the use of an airway adjunct (eg oropharyngeal, or nasopharyngeal). Insertion of an airway adjunct may produce vomiting or laryngospasm.

Ensure the patient is adequately anaesthetized, as laryngoscopy and endotracheal intubation can produce harmful **adrenergic stress response** with adverse increases in pulse and BP. Concurrent iv with esmolol can attenuate this in a dose-dependent way.

medline:11576054

Mechanical ventilation

Indications: Ventilators are used in anaesthesia when there is an operative need for muscle paralysis, or when muscle paralysis is part of a balanced general anaesthetic for a long operation. (Ventilators are used in intensive care for ventilatory support in reversible acute respiratory failure—this is a different topic: see eg p 234.)

Modes:

- Whenever surgeons open the pleural cavity to operate on the heart, lungs, or oesophagus, *intermittent positive pressure ventilation* is required (IPPV). These ventilators have controls to alter the respiratory rate, the tidal volume, and the pressure necessary to inflate the lungs. Other controls may be available to adjust the inspiratory time, the inspiratory flow wave form, the end tidal pause, and the I:E ratio (I:E means the ratio of inspiratory to expiratory time).
- Some ventilators deliver *synchronized intermittent mandatory ventilation* which allows the patient to start breathing spontaneously when paralysis wears off, providing assistance if these breaths are inadequate.
- Specialized patterns of ventilation are not commonly available using ventilators designed for operating theatre use.
- ▶ It is mandatory for ventilators to have disconnect, high pressure, and oxygen failure alarms.

The practical conduct of anaesthesia: II

Intubation This is needed for protection of the airway in:

- Risk of vomiting/aspiration of gastric contents (eg reflux oesophagitis, abdominal disease, major trauma, non-fasted, hiatus hernia, pregnancy).
 - Management of difficult airways. If difficulty is suspected, ensure senior help is available/consider fibreoptic or awake intubation
 - Head and neck surgery, where masks clutter the operative field.
 - Where muscle paralysis facilitates surgery, eg abdominal surgery.
- ▶ Paralysed patients cannot breathe—and so require ventilation.

Intubation requires good muscle relaxation:

- Deep anaesthesia in the spontaneously breathing patient.
- Use of short- or long-acting muscle relaxant.

Short-acting: (suxamethonium, typically 1mg/kg iv) Uses: ● If risk of vomiting ↑. ● When difficulty with intubation is anticipated. ● If the intention is to let the patient breathe spontaneously via the ET tube.

Long-acting: Numerous available, eg vecuronium 0.1mg/kg iv. They take longer to provide suitable conditions for intubation. Remember it will be at least 20min before breathing recommences, so *if there is any doubt as to the ability to maintain a patent airway, then a short-acting (or even no) muscle relaxant is more appropriate.*

Rapid sequence induction Used where the risk of vomiting is high (eg trauma, non-fasted, pregnancy >15 weeks' gestation, hiatus hernia).

- Pre-oxygenate with 100% oxygen for 3min to provide an O₂ reservoir in the lungs for use during the period of induced apnoea.
- Cricoid pressure on induction (firm backward pressure on cricoid cartilage occluding oesophagus may stop gastric reflux to larynx).
- Short-acting muscle relaxant is given immediately after induction. The trachea is intubated, and the cuff of the ET tube inflated.
- Once the ET tube is correctly positioned, cricoid pressure may be released, and a volatile agent added to maintain anaesthesia.

Maintenance of anaesthesia

- 1 Volatile agent added to N₂O/O₂ mixture as before. Either spontaneously breathing or ventilation, with or without opiates. If the patient is ventilated, muscle relaxants are generally used.
- 2 High-dose opiates with mechanical ventilation.
- 3 iv infusion anaesthesia, eg propofol ± opiates. *Maintenance dose of propofol: 4–12mg/kg/h ivi. SE: anaphylaxis; delayed fits (beware day-case surgery).*

Whatever technique is used, the dose and concentration of each agent used is adjusted according to the level of anaesthesia achieved versus the desired level. (This is determined by monitoring vital signs—eg pulse, BP, signs of sympathetic stimulation such as sweating, pupillary dilatation, lacrimation etc.)

End of anaesthesia Change inspired gases to 100% oxygen only, then:

- Discontinue any infusions of anaesthetic drugs.
- Reverse muscle paralysis with neostigmine (dose 2.5mg in adults) + an anticholinergic drug to prevent its muscarinic side-effects (bradycardia, salivation), eg atropine 1.2mg iv (or glycopyrronium—preferred with neostigmine).
- Once spontaneously breathing, put the patient in the recovery position (p 789). Before the ET tube is removed the mouth and oropharynx should be cleared under direct vision. Then administer oxygen by face mask.
- If there are no problems, transfer to the recovery room, but be prepared to reassess the patient at any time.

Intubation technique

Preparation is the key word. Always assess neck mobility, (arthritis, ankylosing spondylitis, goitre etc) pre-op. Beware the obese, short-necked, chinless patient with protruding teeth!

- ET tube sizes (mm internal diameter, ID): 8.5 in adult ♂, 7.5 in ♀.
- Children: ID = $[\text{age in years}/4] + 4.0\text{mm}$ (neonate = 3–3.5mm)
- Length (oral) = $[\text{age}/2] + 12.5\text{cm}$
- Length (nasal) = $[\text{age}/2] + 14.5\text{cm}$

Broselow tape-measures are said to be more accurate. medline:9830856

- Lubricate the tube, and check that its cuff and the laryngoscope work.
- Position the patient with neck flexed and head extended using a pillow ('sniffing the morning air').
- Hold the laryngoscope in the left hand; open the mouth with the right.
- Slide the laryngoscope blade down the right side of the tongue into the vallecula (area between tongue and epiglottis), guarding the lip and teeth with the fingers of your right hand.
- Lift the laryngoscope blade upwards and away from yourself. DO NOT lever on the teeth or you may damage them.
- Lift the epiglottis from view: the cords should become visible. When they are, insert the tube with your right hand.
- Once the cuff of the endotracheal tube is beyond the cords, remove the laryngoscope; ask the assistant to inflate the cuff to prevent air leak.
- Attach to the circuit. Gently inflate lungs. Watch chest move. Do both sides move equally? Is the abdomen moving and not the chest?
- Auscultate both sides of chest. Is air entry equal? Fix the tube with a tie.
- Confirm correct placement with capnography.

► Remember: *if in doubt, take it out*. It is safer to re-intubate than to risk leaving a tube in the oesophagus happily inflating the stomach. Tubes may slip down a main bronchus (usually right). If so, withdraw, until both sides of the chest move equally, and air entry is equal (so avoiding collapse in the unventilated lung, or pneumothorax on the overventilated side).

If you are having problems with intubating: ► Adequate oxygenation is the priority. • Get help • Blind nasal and fiberoptic intubation are options for the experienced • If you find you need another dose of suxamethonium, give atropine first, to prevent bradycardia • Consider a laryngeal mask (below).

Laryngeal mask airway (LMA, a half-way house between a simple airway and an endotracheal tube.) It is used in 50% of elective UK surgery, and in cardiac arrests where a skilled intubator is not present. It consists of a tube with a cuff designed to sit over the larynx. A cuff takes ~30mL of air (depends on size). It is more efficient than masks (eg if arthritis prevents the neck extension needed by masks).

During its use, nasogastric aspiration of air may be needed, but usually only if with used with IPPV. Advantages are that *no laryngoscope is needed* (no damage to teeth or cord stimulation), and *ease of insertion*. Lubricate the cuff, and slide over the palate, so the device sits over the larynx. Learn the technique at the bedside. It is available in a range of sizes from 1.0 to 5.0 (in the UK, available eg via Intavent, tel 01628 594500).

Specialized LMAs can assist with difficult intubation (eg the Fastrach), and allow air and fluid to be aspirated from the stomach (eg the Proseal).

If unable to intubate Learn beforehand where other equipment such as the combitube or cricothyrotomy kit (OHCM p 754) are kept, and how they are used in an emergency.

Monitoring during anaesthesia

Monitoring begins prior to induction of anaesthesia and continues throughout. A **warm, pink, and well-perfused** patient is the aim. Sweating and lacrimation invariably indicate something is wrong: **Respiration**: Rate, depth (IPPV if inadequate). **BP**: Intra-arterial in long cases. **T°**: Particularly important in infants. (Large surface area to body mass may lead to hypothermia.) A warm environment, warming blankets, warming iv fluid, are important in long cases. **Pulse oximetry**: Computes pulse rate and arterial O₂ saturation, but does not obviate the need for manual pulse checks. **ECG**: Reveals rate, arrhythmias, and ischaemia. **CVP**: Helps differentiate hypovolaemia from ↓cardiac function. Insert when large blood loss is anticipated, or in unstable patients. **Capnography** and **Inspired gas analysis** are essential. A low end tidal CO₂ warns of a displaced endotracheal tube, emboli, and more. **Bispectral Index (BIS) EEG monitoring** helps measure of the depth of GA.

Some complications of anaesthesia

► **The commonest respiratory complication is airway obstruction due to loss of muscle tone in soft palate, or 'swallowing the tongue.'**

Atelectasis and pneumonia Atelectasis is best seen on CT (not CXRs). It starts within minutes of induction, and is partly caused by using 100% O₂. In one careful study, it was prevented by using 30% O₂ at induction (just enough to cause adequate oxygenation). Rx: good pain relief aids coughing. Arrange physio + antibiotics (*OHCM* p 162).

Laryngospasm The cords are firmly shut. Treat with 100% oxygen. Attempt to ventilate. It may be necessary to paralyse and intubate.

Bronchospasm Ensure oxygenation. If intubated check tube position (carina stimulation may be the cause: withdraw tube slightly). Check for pneumothorax. (Increase concentration of volatile agent if he is 'light'). Salbutamol 250µg iv, aminophylline (p 271) + hyperventilation with 100% O₂. If persistent, give hydrocortisone 100mg iv and consider epinephrine (=adrenaline, 1–10mL of 1:10,000 iv). **Anaphylaxis** See p 309.

Shivering ± hypothermia Preventable with 'space blankets', and may be treated by radiant heat and pethidine for shivering (0.3mg/kg slowly iv). Be aware that shivering increases O₂ consumption 5-fold.

Scoline apnoea (Abnormal cholinesterase) ventilate until power returns.

Malignant hyperpyrexia (Autosomal dominant). Rapidly rising temperature (>2°C/h) is precipitated by eg halothane or suxamethonium. Masseter spasm may be the first indication, then tachycardia, mottling, hypercarbia, hyperkalaemia, arrhythmias, hypoxaemia, and acidosis.

Treatment: •Get senior help; stop all precipitants and stop surgery •Hyperventilate with O₂ •Active cooling (ice packs, cold iv). •Take to ITU •Dantrolene 1mg/kg iv bolus; repeat as needed till up to 10mg/kg given. •Mannitol for myoglobinuria •Correct acidosis (sodium bicarbonate). •Treat any hyperkalaemia (*OHCM* p 258; children p 280 this book). •Encourage diuresis •Insulin •Steroids, eg hydrocortisone 100mg iv. •Phenotype relatives (muscle biopsy response to halothane and caffeine). •Get a Medicalert® bracelet, and a letter to give to future anaesthetists.

Awareness This is most distressing for patients. Neuromuscular blockade makes its diagnosis difficult; the BIS monitor (above) may ↓incidence. **Prevalence**: ~1% in emergency/obstetric anaesthesia, as doses used are minimized, to avoid shock. It is rare otherwise (eg 0.2–0.4%).

Sedation

Definition: Sedation is an induced reduction in conscious level, during which verbal contact is maintained with the patient.

Doctors in many specialties may be required to administer sedation. The doctor giving the sedation *must not* be also responsible for performing any procedure (such as manipulation of a dislocated joint). His sole responsibility is to ensure that the sedation is adequate, and to monitor the patient's airway, breathing, and circulation. Sedation is not a short cut to avoid formal anaesthesia, and it does not excuse the patient from an appropriate work-up or reasonable fasting. Monitoring is mandatory, and should include pulse oximetry.

It is easy for sedation to become general anaesthesia, with its attendant risks (see p 772). The loss of the 'eyelash reflex' (gentle stroking of the upper eyelashes to produce blinking) is a good guide to the onset of general anaesthesia.

In the elderly, ↓ cognition after GA may persist for months: it is unknown whether lesser forms of anaesthesia can obviate this.

review
medline:11084526

Agents

- Diazepam (as emulsion = Diazemuls®): initial adult dose 2.5–10mg iv.
 - Midazolam: 0.1mg/kg. Initial adult dose 2mg iv over 30sec (1mg if elderly). Further 0.5–1mg iv as needed after 2min. Usual range 2.5–7mg (elderly 1–2mg). SE: psychomotor function ↓.
- In some circumstances (eg manipulation of large joint; painful dressing changes) a narcotic analgesic may be used in addition (eg morphine in 1–2mg aliquots iv, or shorter acting opioids such as fentanyl), or small incremental doses of ketamine may be administered.

review
medline:11228790

Recovery from anaesthesia

- Ensure same monitoring as *during* anaesthesia.
- 40% oxygen via a face mask for >15min to prevent 'diffusion hypoxia'—ie nitrous oxide leaves the blood faster than nitrogen from the air enters it, so greatly diluting the available oxygen in the alveoli.
- Monitor pulse and BP.
- Keep the patient warm.
- Look for hypoventilation—may present as 'see-sawing respiration' (?inadequate reversal—check with nerve stimulator; narcosis—reverse opiates with naloxone—*cautiously* to minimize pain; check for airway obstruction, eg from bleeding tonsil). Ensure adequate analgesia.
- Return the patient to the ward when you are satisfied with his cardiovascular and respiratory status and pain relief.
- Give clear instructions on post-operative fluid regimens, blood transfusions, oxygen therapy, pain relief, and physiotherapy.
- Post-op vomiting is partly preventable by a 5-HT₃ antagonist eg granisetron, or dexamethasone.
- Metoclopramide is less good.
- Data on combining droperidol with 5-HT₃ antagonists are inconclusive.
- Epidural local anaesthetics ↓ GI paralysis compared with systemic or epidural opioids, with comparable pain relief.
- Epidurals may also ↓ post-op risk of respiratory failure.*

meta-analysis
medline:11421822 & 10542894 & 10357347

meta-analysis
medline:11065206

review
medline:11034732

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Local anaesthesia

► All regional techniques may lead to loss of consciousness or loss of airway, and so require the same facilities, expertise, and precautions as for general anaesthesia (eg full resuscitation facilities and patient fasted).

Local anaesthesia is used either alone or to supplement general anaesthesia, the aim is to prevent or reduce nerve conduction of painful impulses to higher centres (via the thalamus), where the perception of pain occurs. (Action is by a membrane-stabilizing effect, impairing membrane permeability to sodium, so blocking impulse propagation.)

Types of local anaesthesia (LA) Amides:

- 1 Lidocaine (=lignocaine) $t_{1/2}$ = 2h; max dose in typical healthy adult = 3mg/kg up to 200mg (500mg with epinephrine, but use with caution).
- 2 Prilocaine $t_{1/2}$ = 2h. Moderate onset. The dose is 3–5mg/kg. (400mg is the maximum dose in adults; 300mg if used with felypressin.) Very low toxicity, so it is the drug of choice for Bier's block (iv regional anaesthesia).
- 3 Bupivacaine $t_{1/2}$ = 3h. Slow onset and prolonged duration. Contraindicated in intravenous regional anaesthesia (Bier's block). (NB: levobupivacaine is a newer less cardiotoxic local analgesic, otherwise similar in action to bupivacaine). For dose see *BNF*.
- 4 Ropivacaine $t_{1/2}$ = 1.8h. Dose: see *BNF*. Less cardiotoxic than bupivacaine. Less motor block when used epidurally. Contraindicated for regional anaesthesia and paracervical block in obstetrics.

Note: 0.5% solution = 5mg/mL. 1% solution = 10mg/mL. So for a 70kg man, the max dose of lidocaine is 20mL of 1% or 10mL of 2% solution. NB: lidocaine injections are less painful if they are warm, or at lower concentrations.

Certain commercially available preparations contain epinephrine (=adrenaline); these should be used with extreme caution, as systemic effects from the adrenaline may arise and prove hazardous, especially in cvs disease or ↑BP.

► Epinephrine (adrenaline) is **ABSOLUTELY contraindicated** in digital or penile blocks, and around the nose or ears. (Ischaemia produced may lead to gangrene.)

Esters (Infrequently used now.)

- 1 Cocaine. Very high toxicity. Short duration of action. Used as a paste preparation for anaesthesia and vasoconstriction prior to nasal intubation or nasal surgery.
- 2 Tetracaine $t_{1/2}$ = 1h. Slow onset. High toxicity. Drops for topical anaesthesia to eye (the eye must be covered with a patch following use), and now topically as an alternative to EMLA®.

Toxicity From excess dose, too rapid absorption, or direct iv injection.

Features: Perioral tingling; numb tongue; anxiety; lightheadedness; tinnitus; seizures; apnoea; collapse; direct myocardial depression; coma.

Treatment: Ensure oxygenation.

Hypotension: try iv fluids first, then consider ephedrine 5mg iv slowly, may be repeated at 3–4min to a maximum of 30mg.

Convulsions: thiopental (=thiopentone) sodium, or benzodiazepines, eg iv Diazemuls® 2.5–5mg boluses. Intubation/IPPV will probably also be needed. cvs collapse: full resuscitation, ie intubation, epinephrine (=adrenaline), atropine, and cardiac massage as required.

Anaphylaxis (p 309) Occurs more commonly with the esters, but can occur with amides. A 2mL test dose may detect possible allergic reactions, and will also warn of inadvertent intravascular injection.

Local anaesthesia: techniques (► Bier's block: see p 700)

► Explain the procedure (co-operation helps). Obtain informed consent.

1 Infiltration Use a small gauge needle. Lidocaine (=lignocaine) 0.5% is the most common agent. Frequent aspiration is important. Remember: after initial injection, subsequent injections through infiltrated areas hurt less. Likewise, infiltration through the cut edges of a laceration is less painful than through the skin.

2 Topical: Tetracaine to eye. Lidocaine 4% spray to cords prior to intubation (nil by mouth for 4h afterwards avoids aspiration).

3 Nerve blocks: Lidocaine (=lignocaine) 1–2%, prilocaine 0.5%, or bupivacaine 0.25%. Nerve stimulation increases success rate. It is now believed that it is the volume of the anaesthetic agent used, rather than the concentration per se, that improves anaesthesia.

Femoral nerve block Direct injection or catheter placement into the femoral sheath. Useful for fractured femur.

- Mark out the inguinal ligament (pubic tubercle to the anterior superior iliac spine). Palpate, and mark, the femoral artery.
- Insert short-bevel needle (insulated if nerve stimulator is used) 1cm lateral to the artery, just below the ligament. A 'click' is felt on traversing the skin, and again on traversing the fascia lata. Find the position either by eliciting paraesthesia, or with a nerve stimulator.
- Aspiration test, then inject eg 30–40mL 0.5% prilocaine.

Spinal anaesthesia

- Insert iv cannula and infuse 300–500mL crystalloid. Check BP.
- Sitting or left lateral position. Surgical scrub; gown & gloves. Prepare back.
- Infiltrate skin with 1–2mL 1% lidocaine (=lignocaine).
- Insert 22–25G spinal needle at L3/4 space (ie below spinal cord). Free flow of csf confirms correct placement. Rotate the needle through 180° to ensure that all the needle aperture is in (avoids patchy blocks).
- Inject 1–3mL 0.5% Marcain Heavy® (bupivacaine+dextrose, a hyperbaric solution that falls by gravity. This allows the height of block to be varied by posture.) Note—much less LA is required during pregnancy.
- Position patient to achieve low, high, or unilateral block.
- Monitor BP—may fall; if so, give crystalloid ± vasopressors (eg ephedrine 5–30mg iv, or phenylephrine 50–100µg).

A small total drug concentration is required—producing sympathetic blockade (vasodilation, hypotension); sensory blockade (numbness); and finally motor blockade (reduction or absence of lower limb power).

Absolute contraindications to spinal anaesthesia:

- Anticoagulant states (risk pressure damage to cord from bleed).
- Local sepsis (risk of introducing infection to csf).
- Shock or hypovolaemic states (effective reduction in circulating volume due to vasodilatation).
- Raised intracranial pressure.

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Relative contraindications to spinal anaesthesia:

- Fixed output states (eg mitral stenosis, aortic stenosis).
- Neurological disease—procedure may be blamed for change in state.
- Ischaemic heart disease.
- Spinal deformity or previous surgery.
- Bowel perforation (theoretical risk of ↑parasympathetic activity, peristalsis and peritoneal soiling).

Extradural (epidural) anaesthesia*

- Insertion of indwelling catheter allows prolonged instillation of LA and/or opiates. Larger volumes of LA are required than with spinal anaesthesia. Lumbar most common site, but cervical/thoracic possible (needs great skill).
- Use aseptic technique, with patient sitting, or in the left lateral position.
 - Do BP. Infuse 300–500mL crystalloid.
 - L3/4 commonest site. Infiltrate with 1–2mL 0.5% lidocaine (=lignocaine).
 - Insert 18G Touhy needle until held firm in ligamentum flavum (~2–3cm).
 - 'Loss of resistance' technique finds epidural space: 10mL 0.9% saline via Touhy needle is difficult to inject while in ligaments, but once in the epidural space, sudden loss of resistance enables easy injection.
 - Fine-bore epidural catheter threaded, needle withdrawn, and catheter placed to needle depth plus 3–5cm.
 - Aspiration test prior to 2mL test done. Wait 3min.
 - Inject required dose, eg 10mL 0.25% plain bupivacaine in 5mL aliquots.
 - Secure catheter in place.
 - Monitor BP every 5min for 15min (slower onset than spinal, therefore hypotension takes longer to be revealed).

Complications of epidural anaesthesia:

- Dural puncture. CSF leak may not be obvious, hence the importance of test dose. (Ward tests for glucose will be +ve if CSF present). Push oral fluids, with caffeine. Nurse flat. Prescribe analgesics for headaches, laxatives to prevent constipation and straining. Blood patch is usually necessary if headache persists longer than 24–48h. *Inform a consultant.*
- Vessel puncture and injection. Treat with oxygen, IV, pressor drugs, atropine if bradycardia (due to block of sympathetic outflow to heart T2–4), and remember the ABC of basic resuscitation.
- Hypoventilation. Due to motor block of intercostals. May or may not require control of ventilation.
- 'Total spinal'—ie injection of a large epidural dose into the CSF. Marked hypotension. Apnoea. Loss of consciousness. Treatment: IPPV. Treat hypotension. ► Death will occur from asphyxia if treatment is not prompt.
- Epidural haematoma or abscess formation: aim for early diagnosis—as this helps prevent big CNS deficits. medline:11153548
- Patch or unilateral block.
- Nerve root damage.

Caudal (sacral epidural)

- Left lateral position.
- Palpate sacral hiatus (~4–5cm above the tip of the coccyx). This is often not easy. Another method is to palpate the posterior superior iliac spines: the line joining them forms the base of an equilateral triangle with the sacral hiatus at the apex.
- Insert 21G block needle perpendicular to skin through the sacrococcygeal membrane, into the sacral canal. A 22G needle may be useful for infants.
- Aspirate, and inject up to 20mL 0.5% bupivacaine in the adult.
- If the injection is difficult (there should be no resistance), or swelling occurs, then the needle is in the wrong place—so **stop!** Withdraw the needle and start again.

Indications:

- Provides anaesthesia for the sacral region.
- Useful, eg in scrotal surgery, low cavity forceps (needs experience because of risk of injecting into baby's head), hernias or haemorrhoids.

*J Rigg 2002 *Lancet* 359 1276 (N=915)

Pain

John Keats, the best known medical student to die of unrequited love, demonstrated that the life of the spirit entails the capacity to feel pain—but, as usual, Nature has been over-generous in endowing us with this capacity—so making the treatment of pain paramount: it promotes wellbeing, sleep, and the honeyed indolence preceding recovery or the easeful passage into oblivion. We tend to think simplistically about pain, assuming that when pain is submitted to us we must respond with something analgesic or anaesthetic—but there are other approaches.*

Remember also that pain relief aids physiotherapy (allowing coughing and mobility), preventing pneumonia.

Pain exacerbates hypo/hyperventilation, hypertension, and tachycardia.

Methods of analgesia (See *OHCM* p 442 and *OHCM* p 426.)

- 1 **Oral:** •Paracetamol 0.5–1g/6h. •NSAIDs: diclofenac 50mg/8h (remember danger of GI bleed; cover with ranitidine or misoprostol; caution in asthma). Effects on renal function are minimal if pre-op U&E is normal. medline:11406042 Tramadol 50–100mg/4–6h PO/IV. Less side-effects than morphine, but less potent. •Opioids, eg morphine sulfate slow release (eg MST-30® or MST-100®/12h PO. NB: most opiates are poorly absorbed from the GI tract). Potent short-acting opioids such as fentanyl, alfentanil, or remifentanyl are used to relieve intraoperative pain, but are used with caution as they cause respiratory depression.
- 2 **Sublingual:** Buprenorphine (synthetic opiate; 'controlled' drug): 0.4mg/6h sublingually, or fentanyl drops.
- 3 **Inhalational:** Nitrous oxide/oxygen (Entonox®), useful for labour pains, changing dressings, and physiotherapy.
- 4 **Intramuscular:** Morphine 0.2mg/kg IM. Pethidine 25–100mg IM is alternative. (Doses for typical healthy adults.)
- 5 **Intravenous:** Boluses or continuous infusion. Opiates (as above). Patient Controlled Analgesic System (PCAS): The patient can give himself boluses, and if this is sufficient, no background infusion of opiates is necessary. Remember to programme maximum dose limit!
- 6 **Regional anaesthesia (RA):** p 776. Epidurals (opiates, or LA, boluses or continuous infusion). Many techniques used (intercostal nerve, brachial plexus, femoral nerve blocks). Coeliac plexus ablation with alcohol may give 6-months' pain relief in upper GI carcinoma. Some meta-analyses of RA for hip surgery find marginal benefits, eg fewer deaths and DVTs. medline:10823094
- 7 **Transcutaneous fentanyl patches.**
- 8 **Transcutaneous nerve stimulation:** (TNS) see p 114.

Chronic pain

- Epidural steroids for chronic backache.
- Nerve blocks: either temporary (LA) or permanent (neurolytic drugs eg alcohol, phenol). Trigeminal nerve, spinal nerve, autonomic blocks (eg stellate ganglion, coeliac plexus).
- Radiofrequency lesions produce localized damage within the nervous system to block conduction.
- Subcutaneous narcotic ± antiemetics continuously, by syringe driver.
- Hypnosis.
- Acupuncture.

Pain in children See p 274 for narcotic and other analgesia in painful conditions such as sickle-cell disease. For burns, see p 688. For post-op relief, see p 198. For pain relief in casualty: see p 678.

Quality in anaesthesia

Deming's definition: quality is meeting customer requirements at a price they are willing to pay. On this view, anaesthesia has various customers rating its quality: surgeons, hospitals, payers, and patients. These groups may have conflicting requirements.

www.cja-jca.org/cgi/content/full/48/1/6

The most important customer is the patient, and their chief requirement is pain relief—but don't focus on this narrowly. Patients are also very keen to have a friendly anaesthetist explaining what to expect. This is the chief area of disparity between what anaesthetists think patients should want and what they actually want. A holistic approach to anaesthesia does not regard these tasks as separate: they are complimentary. Also, avoiding nausea/vomiting and gagging on endotracheal tubes are top priorities whenever these possibilities are explained.

^{n = 147}
medline:10475299

EBM in anaesthesia: possible limitations

In anaesthesia research, surrogate or intermediate outcomes predominate as the end-points which is a weakness when the results serve as the evidence on which to base clinical decisions. More importantly, in interventions requiring skill, dexterity, and dynamic decision-making, do not assume that equally good outcomes are achievable by all, by the simple application of the results of EBM.

medline:9452854

*That some stream of lightning
From the old man in the skies
Can burn out that suffering
No right-taught man denies.

But a coarse old man am I,
I choose the second-best,
I forget it all awhile
Upon a woman's breast.

Daybreak and a candle-end, WB Yeats

12. Pre-hospital immediate care

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Relevant pages in other chapters:

Glasgow Coma Scale (p 680); Child Coma Scale (p 257); cardiorespiratory arrest (p 310–11 & *OHCM* p 764–5); drowning (p 682); burns (p 688); pneumothorax (*OHCM* p 180); asthma (p 270).

Sources The best text we have found is *Pre-hospital Medicine* by Greaves & K Porter, 1999, Arnold, ISBN 0 340 67656 6. *EBM and pre-hospital care*: See F Bunn et al 2001 Report of who pre-hospital care steering committee.

Definition Pre-hospital *Immediate Care* is the provision of skilled medical help at the scene of an accident or medical emergency, or while in transit to hospital. The Diploma in Immediate Medical Care is a benchmark for professional standards in this area (nurses, doctors, and paramedics may apply).

History Baron Dominique Larrey, Napoleon's Surgeon Marshall, may have provided the first skilled help at the scene with his *ambulances volantes* ('flying ambulances'), and was the first to apply the principle of triage—to sort the injured into priorities for treatment (p 786–7).

Note The aim of this chapter is not to provide a full account of the care of any patient prior to hospitalization—this is by-and-large the province of the general practitioner. Rather, our aim here is to highlight those circumstances when doctor and paramedic can work together effectively to save life.

► 'Accidents' are in fact predictable and preventable

Road crashes affect 1 in 4 people in their lifetime. Each day ~3000 people die and 30,000 people are seriously injured on the world's roads. They are the chief cause of death in those <45yrs; most deaths in the 15-19 age group are preventable.

²⁰⁰²
<http://bmj.com/cgi/content/full/324/7346/1107>

In the uk, road deaths have fallen since 1966 despite a >50% ↑ in numbers of vehicles. There are 3500 deaths/yr and ≤45,500 serious injuries/yr in the uk. Data analysis shows little impact on the rate of decline in fatal accidents in the uk from the seat-belt law (1983). One reason may be **risk compensation**—ie safety improvements are transferred by drivers into increased speed or recklessness—illustrating that public health experts must recognise our complex emotional and cognitive reactions to safety issues.

²⁰⁰⁰
<http://bmj.com/cgi/content/full/324/7346/1108> medline:2607298

Air bags also give a complex picture. They reduce fatalities, but front seat children without a seat belt and infants in passenger-side rear-facing car seats are at risk of fatal injury. Burns, high-frequency hearing loss, corneal and retinal injuries also occur.

medline:10703118 &: 11109145

Casualty rates per 100 million vehicle km are similar for cyclists (~200 deaths/yr) and motorcyclists (~500 deaths/yr). Age factors: babies are rarely killed as their **risk exposure** is less. Deaths fall in middle age, rising again before falling in old age. In children, death rates are higher in lower social classes and in more deprived socioeconomic areas.

medline:11144632

Alcohol is a major factor in >10% of preventable crashes. 20% of drivers and 36% of pedestrians killed have blood alcohol levels >the legal limit of 80mg/100mL (17.4mmol/L). This rises to 50% of drivers and 73% of pedestrians killed between 10pm & 4am. There is evidence that older drivers compensate for alcohol-impaired performance by driving carefully; young drivers can compensate in this way, but may choose not to do so (peer group pressure or a delight in risk-taking).

Benzodiazepines, antidepressants, antihistamines, and mobile phone use also contribute to road crashes, and it is important that patients are advised not to drive while on these.

medline:9802269

If a patient has epilepsy or diabetes with hypoglycaemic attacks (or is otherwise unfit to drive, p 466) it is his duty to inform the licensing authorities, and the doctor's duty to request him to do so.

Prevention A vital cognitive shift occurs if the word *accident* is replaced by **preventable occurrence**, implying that accidents are predictable. They typically happen because of laziness, haste, ignorance, bad design, false economy, and failure to apply existing knowledge—and **safety education** can change behaviour.

¹⁹⁹⁶
<http://bmj.com/cgi/content/abstract/324/7346/1129>

Schemes can help young drivers who abuse alcohol, eg the *Driver improvement system for traffic violators*, and re-education by driving instructors. Psychotherapy helps more than lectures. Health education posters picturing tragic consequences to a girl- or boyfriend are successful.

Laws are an effective means of saving lives (eg seat belt and speed laws). Alcohol laws and those governing the roadworthiness of cars presumably prevent accidents, but their effectiveness has not been quantified.

Another effective way of reducing alcohol-related road crashes is to provide good lighting at difficult bends or junctions. Injury in homes may be prevented by such simple measures as child-proof containers, putting holes in polythene bags, using toughened glass throughout the home, and using cooking pans with handles turned in away from toddlers.

Accident statistics (UK)

Every year about 13,000 people are killed in accidents in Great Britain. This includes 1000 children.

UK deaths and other injuries^{1,2}

On the roads

Deaths	3500
Serious injuries	40,000
Less serious injuries	261,400
Children's deaths	270
Children seriously injured	~7000
Deaths in those >65yrs old	1004

Home-based accidents

Deaths	4066
Other injuries	224,000
Injuries in gardens, drives and pathways	388,000

Deaths at work 376

Injuries at work 1,500,000

(~45,000 are serious)

Accidents in the home Old people are particularly at risk. Over half the males and three-quarters of females who die from accidents in their homes were 65 years old or older. About half of those old ladies falling and fracturing their femur will be dead in 6 months (p 708). The commonest cause of accidental death in children is suffocation; in the 15–44 age group it is poisoning, and in the over-45s the chief cause is falls.

Hospital attendances for accidents 7.5 million people/year in the UK attend an accident and emergency department following an 'accident'. 350,000 (4.7%) of these are admitted. One-third of accidents occur in the home, and one-quarter are in those under 5yrs old.

Many more receive treatment from their general practitioner.

Accidents in children

- ~10,000 children are permanently disabled by accidents each year.
- Accidents cause 1 child in 5 to attend the A&E department each year.
- Accidents are the commonest cause of death among children aged 1–14yrs, and they cause half of all deaths in those aged 10–14yrs.
- School age children (4–14yrs) in road crashes restrained with a seat belt are 2–10 times as safe as unbelted children, and were at least as well protected as adults wearing seat belts.¹

Useful contacts and addresses:

- British Safety Council, 62 Chancellors Road, London W6 9RS (UK).
- Construction Health and Safety Group, St Ann's Road, Chertsey, Surrey KT16 9AT (UK); tel. 020 8741 1231.
- Scottish Chamber of Safety, Heriot-Watt University, Riccarton, Currie, Edinburgh EH14 4AS (UK).
- Injury Prevention (full-text) http://ip.bmjournals.com/current.shtml#ORIGINAL_ARTICLES

¹ H Binns 2002 *BMJ* i 927 and A Mickalide 2002 <http://bmj.com/cgi/content/full/324/7346/1108>

² Royal Society for the Prevention of Accidents (RoSPA), 353 Bristol Rd, Birmingham B5 7ST (tel. UK 0121 248 2000)

³ APLS group Advanced Paediatric Life Support, BMA

►► Basic life support (BLS)

Synonyms Artificial Respiration; cardiopulmonary resuscitation (CPR).

Definition BLS is the provision of life support—expired air (your own) ventilation + external chest compression, without any equipment.

SAFE approach

- As you approach the patient shout for help (pointing to an individual if possible, to activate him or her).
- Approach him with care—are there any hazards to yourself (p 790)?
- Free the patient from immediate danger.
- Evaluate the patient's 'ABC' (see below).

The SAFE approach

Shout for help

Approach with care

Free from danger

Evaluate ABC (Airway, Breathing, Circulation)

Establish unresponsiveness Shake gently by the shoulder while stabilizing the forehead with the other hand. Ask 'Are you all right?' If he responds, put in the recovery position (p 789)—if not, check for breathing.

Breathing Open and clear the airway (finger sweep; remove denture plate or loose false teeth—leave a well-fitting set, otherwise the mouth collapses making expired air ventilation difficult). If breathing, put in the recovery position; if not, go to get help now. On return, open the airway, and give 2 rescue breaths (slow inflations, just enough to make the chest rise—and achieve a tidal volume of ~400–500mL).

Circulation Feel the carotid pulse for up to 10s (can be difficult, and other signs of circulation may also be used). If felt, give 10 rescue breaths, then go for help. If no pulse, give 15 chest compressions (lower $\frac{1}{3}$ of sternum, 2 fingers' breadth above xiphisternum); depress by $\frac{1}{3}$ of the chest's diameter, a rate of ~100/min).

Children (1–8yrs) Same sequence, but use one hand one finger's breadth above xiphisternum (rate 100/min). Avoid blind finger sweep—may impact foreign body in conical upper airway; do look into the airway for easily removable foreign body. See p 310.

Infants (<1yr) If not breathing, give 5 rescue breaths, then check pulse (use brachial pulse as the neck is very short). If no pulse, give 20 cycles of 1:5 ventilations-to-compressions (compression rate 100/min) then go for help—take the baby with you and continue BLS while phoning for help. Avoid blind finger sweeps.

► See p 310 for the algorithm of the European guidelines for basic and advanced life support.

►► Heimlich manoeuvre

Respiratory obstruction by choking on a foreign body is not an infrequent cause of death—which may be prevented by performing the Heimlich manoeuvre. If the person who is choking is conscious and standing, first ask him to bend forward and cough. If this fails, get behind him, and with your arms encircle his abdomen, mid-way between umbilicus and xiphoid process. One hand makes a fist, positioned thumb-to-abdomen. The other hand grasps this fist, and with a sharp movement presses it up and into the abdomen, to dislodge the foreign body. If the victim is already comatose, this manoeuvre can be accomplished with him lying on his back—using the heel of the hand to press with, rather than a fist. Repeated thrusts may be needed. In adults, sweep the mouth with a finger to remove the foreign body.

► Do not perform this manoeuvre on small children, instead use back blows (below) and chest thrusts (similar to cardiac compressions).

Complications (Rare) If applied incorrectly, direct trauma to abdominal viscera may result \pm thrombosis of an abdominal aortic aneurysm or dislodgement of thrombus causing bilateral acute leg ischaemia.¹

Back blows These are no better, and no worse, than the Heimlich manoeuvre in relieving foreign body obstruction. Neither should be taught or practised to the exclusion of the other. Alternate 5 back blows with 5 abdominal/chest thrusts. Use the heel of the hand to strike forcefully between the scapulae, with the patient leaning forwards.

¹ L Forbes 2002 *Ann Vasc Surg* 16 130 [11904818]

Road crashes*—1: assessment and triage

If you are the first on the scene, the following page (which assumes a highly organized response) will seem impossible on a dark night, alone. So the first priority is to get help. You may be surprised in how short a time it all becomes organized to give the picture described below. Requesting the fire service when dialing emergency services may be the quickest way to get a dozen trained first-aiders to the scene with unrivalled skill in extrication.

The distribution of trauma deaths is trimodal—*immediate* (aortic deceleration injury; severe head injury), *early* (hypoxia and hypovolaemia), and *delayed* (sepsis; multiple organ failure). Prevention (speed restriction; road lighting; seat-belt laws; no drink-driving; air bags) is better than cure—and medical intervention is too late for the immediate deaths.

Priorities at the scene These are command, safety, communications, assessment, triage, treatment, and evacuation.

Command: Once in attendance the doctor is responsible for the provision of all medical care. He will work closely with the ambulance service, but will have supplementary skills (eg providing potent iv analgesia).

Safety: Yourself First ensure your own safety (do not approach fire or chemical hazards (p 790) until the fire service have made the area safe; wear high-visibility clothing, and carry gloves/eye protection/ear defenders). **The scene** If first to arrive, park obliquely behind the incident ('fend off') and leave hazard lights/green beacon on. **The casualties** remove from any immediate danger; protect from further injury during extrication (eg cover with blanket when windows broken). ►Check for a hidden victim, eg under a car or over a wall.

Communications: Liaise with police (they are in overall command), fire service (for any hazards; they can remove the casualty quickly if he is about to die, or in a very controlled manner, eg if an isolated spinal injury), and ambulancemen (identify if they have extended skills; use appropriately). Speak to the receiving hospital by radio or telephone (p 794, radio procedure), and relay the number and severity of casualties.

Assessment: Relate vehicle damage to potential injuries in the casualty ('reading the wreckage'). Is the casualty trapped—relative (cannot move a broken arm to open the door), or absolute (eg feet caught in pedals)?

Triage: From the French *trier*, to sift or sort (coffee beans), this process sorts casualties into priorities for treatment. Divide into **Immediate** (colour-code RED, will die in a few minutes if no treatment, eg obstructed airway, tension pneumothorax); **Urgent** (YELLOW, may die in 1–2h if no treatment, eg hypovolaemia); and **Delayed** (GREEN, can wait, eg minor fractures). Those who will certainly die are labelled **Expectant**—to treat them may delay you helping the salvageable, who then die unnecessarily (BLUE, but not all triage labels have this colour—use GREEN). Do not forget to label the dead (WHITE or BLACK), otherwise emergency personnel may repeatedly take a doctor to the same victim, so wasting time and resources. Note that alcohol or drug abuse (eg cannabis medline:10923130 & 7626673) which may have caused the accident in the first place may make assessing of casualties much harder.

Note that triage is dynamic. It starts with a brief-look assessment (see opposite), but later may involve a detailed examination. Priorities (and label colour) will change while awaiting, and after treatment.

*I Roberts 2002 *BMJ* i 402—RTA (road traffic accident) is a misnomer: most result from speed and carelessness, and are all too predictable, rather than being truly accidental.

How to triage¹

This is a simple system: its main virtue is *speed*. All casualties should be reassessed when time and resources allow. Go through the following sequence until you arrive at a triage priority (in CAPITALS below)—assign its colour label to the casualty and move immediately to the next. Do NOT stop to treat, or you will surrender control of the incident.

1 • Can the patient walk?

Yes → DELAYED (Code with green label)

No → check for breathing. . .

2 • Is the patient breathing?

No → open the airway

Are they breathing now?

Yes → IMMEDIATE (Red)

No → DEAD (White)

Yes → count or estimate the rate (eg over 15sec)

<10 to >30 per minute → IMMEDIATE (Red)

10–30 per minute → check the circulation. . .

3 • Check the circulation

Capillary refill >2sec (or pulse >120) → IMMEDIATE (Red)

Capillary refill <2sec (or pulse <120) → URGENT (Yellow)

¹ Advanced Life Support Group *Major Incident Medical Management & Support: the Practical Approach*, London 1995, BMA Publishing—also *ATLS 1997 Manual* 6e, p409 ISBN 1-880696-10-X

Equipment

In your car boot carry the following minimum equipment:

Oropharyngeal airway(s)	Dressings	Bandages
iv cannulae (14G+16G)	iv fluid (eg Haemaccel®)	Giving set
Sticky tape	Scissors + Torch	Fire extinguisher

You should give a high priority to making yourself safe and visible, eg: reflective jacket, hard hat, Wellington boots.

Extra equipment Cervical collars; Mini-Trach II® (cricothyrotomy kit); chest drain set; sutures; local anaesthetic; analgesia; splints; stethoscope (more a badge of office than a useful tool owing to noise).

►► First aid treatment

A: Airway: Remove false teeth and any vomit. Lie in the lateral position. If trapped in a car, continue to manually stabilize head and neck when a rigid collar is applied. Extricate on to a spinal board, and immobilize the head and neck with foam headblocks and securing straps.

B: Breathing: If spontaneous, give O₂ at 15L/min via tight fitting face mask with reservoir. If not breathing, ventilate mouth-to-mouth. medline:11098945 If available give 100% O₂ via bag-valve-mask and intubate the trachea.* If **tension pneumothorax** suspected (cyanosis, engorged neck veins, shifted trachea), pierce chest with a large iv cannula in 2nd interspace in the midclavicular line, on the side from which the trachea is deviated. If there is an **open chest wound**, cover with an Asherman chest seal (medline:10420335 adhesive disc with central rubber flutter valve).

C: Circulation: Is there a pulse? If not, start external cardiac massage. (Survival from traumatic cardiac arrest with no vital signs at the accident scene approaches 0%.) If there is bleeding, this will almost always be stopped by pressure and elevation of the part. Avoid tourniquets these are often forgotten and left on too long. Insert 2 large iv cannulae (eg antecubital fossae 14G or 16G; secure these well and splint the arm, eg Armlok®) and start fluid resuscitation (eg Haemaccel®). A policeman or fireman makes a willing drip-stand. Ensure that the cannula is not pulled out when the patient is moved. Suspected internal bleeding and fractures of long bones indicate that iv fluid is needed, as blood loss may be considerable (fractured pelvis 2L, femur 1L, tibia ankle and fractures of several ribs ~½L each). Splinting (eg one leg to the other) helps reduce blood loss as well as relieving pain. NB: the ATLS target is definitive haemorrhage control, rather than plasma volume normalization.

D: Disability: Assess responsiveness rapidly with AVPU:

- A = Alert
- V = responds to Vocal stimuli
- P = responds to Pain
- U = Unresponsive

E: Exposure = head-to-toe exam (secondary survey), but is often not practical outside hospital. Reassess ABC; quantify coma level (adult Glasgow Coma Scale, p 680; child, p 257). Remember to give analgesia (p 792).

Transport: Keep the injured warm. Procedures can be performed in moving ambulances (eg iv cannulation) if the patient can be extricated quickly.

*Rapid sequence intubation (RSI) is as do-able in the field and in hospital [@11033272]



The above is one way of positioning the unconscious so vomit is not inhaled and the cervical spine is kept supported in the midline—minimizing risk of spinal cord injury. It helps if there are 2–3 people to 'log-roll' the casualty into this position, so that the neck is never out of alignment with the body. There have been reports of neurovascular risk to the arm supporting the head, so other positions are being tried, but none has been formally validated.

'Scoop and run'—or 'stay and play'?

'Scoop and run' refers to rapid evacuation of casualties to a trauma centre (or A&E department); 'stay and play' entails detailed resuscitation at the scene of trauma. In deciding which is better, take the following into account.

- There is no single answer for all circumstances. Local geography plays a part, as does the fitness of the casualty. Only experience can tell which option is most appropriate—and luck may well play its part too.
- 'Scoop and run' and 'stay and play' are two ends of a spectrum of activity. Often a minimal amount of resuscitation can be done without causing too much delay—ie: get the best of both worlds. medline:9133174
- Penetrating chest injuries have been studied in a pseudo-randomized trial (N=289; full randomization was impossible but 'scoop and run' operated for one day, and 'stay and play' for the next). This showed that the balance of benefit probably lies in 'scoop and run' for this type of injury. Fluid resuscitation aiming for normotension without an operating theatre to hand may be dangerous, as clot may be displaced by the rising BP—with subsequent fatal haemorrhage, which could only have been prevented by major surgery. In this study, 70% in the delayed-resuscitation group survived, compared with 62% who received immediate fluid resuscitation. Duration of hospital stay was shorter for survivors in the delayed-resuscitation group. But note that the results only achieved significance when pre-op and post-op deaths were pooled, and that rates of complications were similar in the two groups (eg respiratory distress syndrome, coagulopathy, wound infection, and pneumonia). medline—7845437 & 7935634

Road crashes—2: hazards and extrication

Hazards

Fire: <5% of road traffic crashes result in fire, and <1:500 result in significant burns. The world's worst ever pile-up was in the Salang tunnel, Afghanistan, in November 1982, involved a petrol tanker explosion with an estimated 1100–2700 killed.

Electricity: Power shorts are common, eg from bird strike, and power may be restored after 20 min without investigation—you must phone the power company to ensure the source is turned off. High tension cables can be lethal even when standing several metres away.

Rail: An electrified rail may be short circuited by a bar carried by the fire service, or the operating rail authority. Remember that cutting power does not stop diesel locomotives that may also operate on the same line: trains may be stopped by signal lights, red flags, or a series of charges placed on the rail—the noise warns the driver.

Chemical: Lorries carrying hazardous loads are required to display an orange 'HAZCHEM' board (see opposite). This contains information on how to fight a fire, what protective equipment to wear, if the chemicals can be safely washed down the storm drains, and whether to evacuate the area (TOP LEFT); a United Nations (UN) product identification number of four digits (MIDDLE LEFT)—eg 1270 = petrol; a pictorial hazard diamond warning (TOP RIGHT); and an emergency contact number (BOTTOM). A white plate means the load is non-toxic. The European 'Kemler' plate contains only the UN product number (BOTTOM) and a numerical hazard code (TOP—note repeated number means intensified hazard).

As a concession to freight carriers, mixed loads of <500kg need only be identified by a plain orange square at the front and rear.

To obtain information about the chemical at the scene of an accident, look at the transport emergency card (TREM card) carried in the driver's cab; the fire service will be linked with CHEMDATA—a computer database at the national chemical information centre at Harwell; alternatively phone Poison's Information Centre (eg 0207 635 9191), or the company.

► Do not approach a chemical incident until declared safe by the fire service.

Principles of extrication

- Stabilize the vehicle where it lies—movement may exacerbate injury.
- Make the vehicle safe—switch off ignition; immobilize the battery; switch away any petrol.
- Identify the time-critical patient—some will die unless rapidly removed from the vehicle, at whatever cost.
- Read the wreckage—relate the damage of the vehicle to potential injuries: steering wheel deformed = chest injury; dashboard intrusion = patella/femur fracture ± posterior dislocation of the hip. Bodies are softer than metal: major bodywork distortion = major injury.
- The easiest way to enter a car is through the door—try this before removing the windscreen or the roof!
- Remove the wreckage from the casualty, not the casualty from the wreckage; don't try to manoeuvre the casualty through too small a hole.
- Don't move from one entrapment situation straight into another—necessary spend a short time stabilizing the patient before moving into the back of the ambulance.

<p>Hazchem advice for the control of the chemical</p>	
<p>Substance identification number and name</p>	<p>4WE 2513 Bromoacetyl bromide</p>
<p>Specialist advice and telephone number</p>	<p>0705 826435</p>
<p>Suppliers name (optional)</p>	<p>PACE TRANSPORT</p>

Hazard symbol and description in a diamond on white square background (rest of label is black lettering on an orange background)

Danger labels



Pre-hospital analgesia

Why should I give pain relief? There are more than humanitarian reasons for giving analgesia. Catecholamines released with pain may further reduce peripheral perfusion and oxygen delivery in hypovolaemic shock, or increase myocardial oxygen demand following myocardial infarction.

When do I give pain relief? Unless all hope of life and rescue has been abandoned, the priorities of securing an airway and stabilizing the cervical spine, maintaining ventilation, and optimizing the circulation always come before analgesia. The effects of any drugs must be weighed against their potential side-effects (respiratory depression; sedation).

How do I relieve pain? *Psychological:* Beecher noted in 1944 at Anzio* that soldiers were indifferent to serious injury. This is unlikely in road crashes; a soldier is released from war horrors by his injuries, but a crash victim is just beginning his nightmare. Reassurance that 'the doctor is here' is important.

Splintage: Simple splints can be improvised from clothing; an uninjured leg can splint the injured one. Inflatable air splints are not very robust, although are light and easy to apply. Box splints (fold around the limb) are popular but poorly accommodate a deformed limb, when a vacuum splint (full of tiny beads) is better. Traction splints should be used for a fractured femur: they also reduce blood loss, morbidity, and mortality (the Thomas splint reduced mortality of open fractured femur from 80% to 20% in World War I).

Gaseous: Nitrous oxide provides comparable analgesia to 10mg morphine. It is mixed with 50% O₂ as Entonox® or Nitronox® in blue cylinders with a white top. It separates at -6°C (O₂ on top). **Do not** use if Caisson disease suspected ('the bends', p 802), or pneumothorax (may tension).

Opioids: Morphine is the gold standard. Give in small aliquots (1–2mg) iv. Don't give im if poor peripheral perfusion might result in erratic absorption. Naloxone (0.4–1.2mg iv) must be available at all times. Nalbuphine is a synthetic prescription-only opioid: it is increasingly used by paramedics. Nalbuphine dose: small aliquots, up to 10–20mg slowly iv, repeated after 30min if needed; for a child the max dose is 0.3mg/kg. As a partial antagonist, it will compete with any morphine given later.

medline:9866242

NSAIDs: IM NSAIDs (diclofenac 75mg IM, repeated once only, at 30min if needed, or ketorolac 10mg IM stat then 10–30mg/2–6h as needed, max 60–90mg/day) has a rôle in musculoskeletal pain—and it doesn't entail monitoring for sedative effects; it isn't recommended for children. medline:9110007 Ketorolac can be given iv over ≥15sec; avoid in: hypovolaemia; labour; asthma.

Ketamine: This is a potent, short-acting analgesic at 0.25–0.5mg/kg iv. At higher doses it can be used as sole anaesthetic agent (2mg/kg iv will produce 5–10min of surgical anaesthesia); 'emergence delirium' is common unless a benzodiazepine (midazolam ~2mg iv) is also given. medline:10989856

Sedation: An anxious or aggressive patient is often in pain or hypoxic. Reassurance, a clear airway with supplemental oxygen and analgesia is better than sedation, although this is needed rarely: give small aliquots of midazolam (p 773, up to 1.5mg in elderly—have flumazenil to hand).

Local anaesthesia: Peripheral nerve blocks aid release of a trapped limb. Femoral nerve block is most used and provides complete analgesia (anaesthesia) for femoral fractures (less effective for low shaft fracture). Locate the artery in the groin and put 10–20mL 1% plain lidocaine (=lignocaine) in a fan shape lateral to the artery. Aspirate frequently to avoid intravascular injection. Maximum dose <3mg/kg or 0.28mL/kg of 1%.

*Anzio, 33 miles south of Rome, was a crucial Allied beachhead in the recapturing of that 'Eternal City' (5/6/44)

Communications

The radio net A radio user is identified by a 'call sign'. Messages are usually passed through a central controller (call sign 'control', or 'zero') without being able to hear other users—'two frequency simplex'; but on some nets all users can hear and talk directly to each other—'single frequency simplex'.

Acquiring a 'radio voice' This takes practice. *Rhythm* should be steady; *speed* is slower than normal speech; *volume*—do not shout or whisper; *pitch* should be raised if the voice is gruff. Remember: 'RSVP'.

Using a radio Switch on and check battery light (switch off to change battery). Listen (single frequency) or look at the 'channel busy' light (two frequency)—you do not want to interrupt any message. Wait 1–2sec after pressing the transmit button before speaking, or the important first few words are lost. Release the transmit button after speaking, or you will prevent others from transmitting. VHF radios have a longer range than UHF.

Messaging To *initiate* say the receiver's call sign then your own. Say 'over' to indicate when the receiver should reply. To *continue* a message always start with your own call sign when you speak. To *end* a transmission say 'out'.

Example: 'Zero from Mike One, message over.'

'Zero, go ahead over.'

'Mike One, moving now to new location out.'

Remember that anyone can be listening; don't be a comic ('Send the rover over, over'); don't swear; address by appointment rather than name.

Key words Spell long or difficult words using the NATO phonetic alphabet (see opposite)—you do not want an 'empty box' when you asked for 'Entonox'. Long numbers (drug dose; grid reference) are given whole, then digit by digit (eg 1000 = 'one thousand, figures one-zero-zero-zero'). 'Roger' or 'OK' means you have understood; 'say again' means repeat the message ('Repeat' is the order for artillery to fire again and is avoided, at least on military networks!). ETA/ETD are common abbreviations for estimated time of arrival/departure. Avoid radio gibberish ('roger dodger/ten four/over and out'; 'negative' and 'positive' for 'yes' and 'no').

Telephone Mobile telephones are useful for pre-hospital communication. However, in major incidents all cells are rapidly utilised (often by the press). In the UK, on application to the Cabinet Office the telephone may be registered to operate on a number of restricted cells in cases where the system is overloaded—this is ACCess OverLoad Control (ACCOLC).

Rank structure

<i>Epaulette</i>	<i>Police</i>	<i>Fire</i>	<i>Ambulance</i>
2 pips	Inspector	Station Officer	Officer band 1
3 pips	Chief inspector	Asst. Divisional Officer	Officer band 2
Crown	Superintendent		
Laurel 'U'		Asst. Senior Officer	
Laurel 'U' around pip		Divisional Officer	

UK fire service helmets:

<i>Colour</i>	<i>Black bands</i>	<i>Rank</i>
Yellow	Nil	Firefighter
Yellow	One thin	Leading fireman
Yellow	Two thin	Sub-officer
White	One thin	Station Officer
White	One thick	Assistant Divisional Officer
White	One thick, one thin	Divisional Officer

The NATO phonetic alphabet

ALPHA	BRAVO	CHARLIE	DELTA
ECHO	FOXTROT	GOLF	HOTEL
INDIA	JULIET	KILO	LIMA
MIKE	NOVEMBER	OSCAR	PAPA
QUEBEC	ROMEO	SIERRA	TANGO
UNIFORM	VICTOR	WHISKEY	XRAY
YANKEE	ZULU		

Radio procedure—number pronunciation

- 1 WUN
- 2 TOO
- 3 THUREE
- 4 FOWER
- 5 FIYIV
- 6 SIX
- 7 SEVEN
- 8 ATE
- 9 NINER

►►The major incident

Planning For a hospital to be prepared to cope with multiple casualties there must be planning. Each hospital will produce a detailed *Major Incident Plan*, but additionally the tasks of key personnel can be distributed on individual *Action Cards*.

At the scene A medical incident officer (MIO) will be requested from Hospital or **BASICS** scheme (British Association for Immediate Care—usually GPs). Mobile medical teams (eg A&E senior doctor/surgeon, anaesthetist + 2 to 4 nurses) should come from hospitals not accepting the main casualties. **BASICS** doctors should be requested to the scene by radio via the ambulance station). Further **BASICS** doctors make valuable treatment officers, as they arrive.

Safety: Is paramount—your own and others. Be visible (luminous monogrammed jacket) and wear protective clothing where appropriate (safety helmet; waterproofs; boots; respirator in chemical environment).

Triage: See p 786.

Communications: Are essential—and frequently an area where improvements are needed. medline:9133172 The police are in overall control of the scene. Each emergency service will dispatch a control vehicle and will have a designated incident officer for liaison. Support medical staff from hospital report to the medical incident officer: his job is to assess then communicate to the receiving hospital the number and severity of casualties, to assess need for further medical teams, to oversee triage and treatment (with the Ambulance Incident Officer). He must resist temptation to treat casualties as this compromises his rôle.

Equipment: Must be portable (in small cases/backpacks) and include: triage labels, intubation* and cricothyrotomy equipment; intravenous fluids (colloid); bandages and dressings; chest drain (plus flutter valve); amputation kit (when required ideally two doctors should concur); drugs—*analgesic*: morphine; *anaesthetic*: ketamine (p 792); *specific antidote* if a chemical threat; cardiac resuscitation drugs; drugs to cover common medical emergencies: eg GTN spray, salbutamol inhaler; limb splints; defibrillator/monitor; \pm pulse oximeter; 'comfort bag' for staff and others (£1 coins, sweets, toilet paper, etc).

Evacuation: Remember: with immediate treatment on scene, the priority for evacuation may be reduced (eg a tension pneumothorax—**RED**—relieved can wait for evacuation—becomes **YELLOW**), but those who may suffer by delay at the scene must go first (eg unconscious closed head injury; myocardial infarct). Send any severed limbs to the same hospital as the patient, if possible keeping them on ice (not *in* ice as freezing harms tissues).

At the hospital a 'major incident' is declared (eg if >10 serious injuries). A control room is established and the medical coordinator ensures staff have been summoned, nominates a triage officer and supervises the best use of inpatient beds, intensive care, and theatre resources. When the incident is declared clear of casualties, the major incident may still continue for some time at the hospital. medline:9133172

*It is wise to include a Broselow tape-measure for giving child tube sizes relative to height—more reliable than (age in years +16) divided by 4. medline 9830854

When do major incidents become complex emergencies?

- If the context involves administrative, political, or economic anarchy.
- If the incident sparks a self-perpetuating chain of violence.
- If the incident is not a random event, but focused on one ethnic group.
- If competition for wholly inadequate resources compounds their inadequacy.
- If the incident leads to displacement of children.
- If the incident promotes a state of war.

www.txt.co.uk/full.cfm?1032

Helicopter transport

►It is often better to spend 30min transporting serious injuries to a well-resourced trauma centre with consultants standing by, than to spend 10min transporting such a person to a small hospital where the most skilled help is not *immediately* available. If the small hospital has to transfer the casualty to a trauma centre, no significant savings may have been made compared with immediate helicopter transfer to the centre. medline:10729675

The importance of helicopters for casualty rescue/transport is increasingly recognized—but be aware of limitations. Helicopters may be used for transporting casualties to hospital, or for interhospital transfer. **Advantages:** Speed over long distances; access to remote areas; delivery of highly trained doctors and special equipment to the scene—eg ready to intubate, paralyse and ventilate, and give mannitol ivi if head injury. medline:8637337 **Disadvantages:**

- Mid-air crashes in air ambulances have occurred, and are hard to survive.
- Noise and general stress, leading to anxiety and disorientation, and hampering communication—reassure and provide with a headset.
- Vibration exacerbating bleeding and pain from fracture sites.
- Cold—beware in those hypothermic rescues from sea or mountain.
- Problems related to altitude; aircraft limitations, eg weather, landing site, limited carriage space (especially if additional medical personnel).
- Police craft don't allow ECG monitor/oximeter due to magnetic radiation.
- Many published reports of the advantages of air ambulances lack rigour.

The gains of helicopter transfer depend on how many severe injuries occur. One UK study concluded that only ~13 lives would be saved per year in London if it was reserved for the severest cases (ISS >15, p 800) in lesser trauma, there is evidence that outcome is less good. medline:7627033

Helicopter safety •Always approach from the front of the aircraft, in full view of the pilot. Secure loose items, eg headgear.

- Do not enter/leave the rotor disc area without permission (thumbs up signal from pilot). Lower your head in the rotor disc area.
- Do not touch the winch strop/cable until the earthing lead has contacted the ground. Also, be sure to avoid the tail rotor.
- Make sure no-one is smoking within 50m of the aircraft.

Problems of altitude Hypoxia is unlikely unless there is cardiac or lung disease, anaemia, shock or chest trauma, as helicopters rarely fly high enough to produce a significant fall in P_aO_2 .

Reduction in atmospheric pressure results in an expansion of enclosed gases on ascent. This produces pain in blocked sinuses, expansion of a pneumothorax, abdominal wound dehiscence (avoid flying for 10 days post surgery if possible) and renewed bleeding from a peptic ulcer. Remember drips may slow down.

On descent, beware of endotracheal tube cuffs and military antishock trousers (MAST*) deflating significantly (particularly if applied at altitude eg on hillside). Rapid descent may induce barotrauma.

Specific problems •Decompression sickness (p 802): if air is breathed under pressure (divers), nitrogen dissolves in blood and tissues. On rapid ascent after a dive the nitrogen will come out of solution as bubbles, producing joint pains ('the bends') ± urticaria, CNS defects and shortness of breath. ►Do not fly if dived <30m within last 12h or >30m within 24h.

- Ischaemic chest pain or infarction is not a contraindication to flying.
- Psychiatric illness (eg mania) may preclude safe air transport.
- Burns >20% need preflight nasogastric tube insertion (prevents gas expansion of an ileus) ± in-flight pressure-controlled ventilation. medline:9169941

*MAST (=medical anti-shock trousers)

MAST inflation may impair breathing & ventricular function. medline:10349920 They have proved deleterious in those with moderate hypotension (systolic BP 50–90mmHg) who face only a short ride to a hospital, especially those with thoracic injuries. MAST's rôle in severe shock or long pre-hospital transport times remains unclear. In severe shock, improvement in BP and oxygenation to the heart and CNS may override any negative effects of continued haemorrhage. medline:10787852

Trauma scoring

Essence Trauma scoring can be used at the roadside to predict the probability of survival (Ps), and thus the severity of injury, which may influence where to take the patient (nearby district general hospital, regional hospital, or Trauma Centre?). Retrospectively, physiological data may be combined with an anatomical injury score to compare performance (expected vs actual survivors) between hospitals. NB: trauma scoring in children is problematic. See expert literature.*

Trauma Score (TS): The original score introduced in 1981 in USA to allow paramedics to make an objective decision on whether patient needed Trauma Centre facilities. Measures respiratory rate + effort; systolic BP; capillary refill; Glasgow Coma Scale (GCS). Maximum score 16. Score ≤ 13 means mortality $\geq 10\%$ —USA take to Level 1 Trauma Centre; UK take to large hospital and alert trauma team en route.

Revised Trauma Score (RTS): Measures respiratory rate, systolic BP, and GCS only: other parameters were found to be poorly reproducible on analysis of a large North American database. Each parameter has values coded to give score of 0–4. These scores are multiplied by a weighting coefficient, then added together to produce the RTS—it is not a suitable roadside tool. RTS has a more reliable correlation with Ps than does TS.

Triage Revised Trauma Score (TRTS): This uses the same coded values of respiratory rate, systolic BP, and GCS from the RTS, but a fall by one point in any parameter is taken as significant (thus TRTS ≤ 11 is significant). This is the most useful pre-hospital trauma triage tool.

Limitations: These scoring systems are not validated for the very young or elderly; up to 20% may be underscored on their initial assessment (if attended rapidly, before physiological decompensation).

Injury Severity Score (ISS) TS, RTS, and TRTS use physiological variables to predict Ps. Anatomical injury (data from operation or postmortem notes) can also be used—injuries are scored from 1 (minor) to 6 (fatal) using tables from the Abbreviated Injury Scale (AIS). The body is divided into 6 regions and the 3 highest scores from different regions are squared and added. Maximum score is 75 (5 squared \times 3) since AIS 6 in any body region is fatal—and therefore awarded a score of 75. ISS correlates closely with Ps. ISS ≥ 16 implies mortality $\geq 10\%$ and is termed 'major trauma'.

Limitations: Injuries can be difficult to code; isolated head injury with AIS of 3 has high mortality, but is excluded from 'major trauma' outcome analysis.

TRISS methodology Trauma audit programmes are established in USA & UK—*Major Trauma Outcome Study (MTOS)*. medline:10821308 TRISS is a complex formula incorporating RTS, ISS, patient age, and whether blunt or penetrating trauma. It is poor at predicting individual outcome, but allows comparison of overall performance between hospitals, or the same hospital following, say, introduction of trauma teams. ASCOT (*a severity characterization of trauma*) is newer but not necessarily better. medline:10804601

Z & M statistics compare outcome in different populations. Z measures the difference between the actual and predicted number of deaths. M (the 'injury severity match') compares the range of injury severity to the main database—if $M < 0.88$, Z is invalid. medline:11142269 &: 9624342

*D Potoka 2001 J Ped Surg 36 106 See also [11150447] [11038093] [10389962]

Blast injury

Blast injury may be encountered in domestic (eg gas explosion) or industrial (eg mining) accidents, or as the result of a terrorist bomb. Terrorism worldwide is responsible for 15,000 injured or killed in the last 20 years and most casualties are from bombs. Death may occur without any obvious external injury, often due to air emboli, the correct cause first being recognized by Pierre Jars in 1758 as a 'dilatation d'air' (ie blast wave). Explosions cause injury in six ways:

1 Blast wave A transient (milliseconds) wave of overpressure expands rapidly away from the point of explosion, its intensity inversely proportional to the distance cubed. It produces: (a) cellular disruption at air-tissue interface ('spalling'), ie perforated ear-drum at 100kPa, 'blast lung' at 175kPa; (b) shearing forces along tissue planes: submucosal/sub-serosal haemorrhage; (c) re-expansion of compressed trapped gas: bowel perforation, fatal air embolism (coronary artery or cerebral).

Blast lung is often delayed (up to 48h). It is **rare** in survivors—only 0.6%. Suspect it if there is a perforated drum, but this is **not** a prerequisite (as position of drum in relation to blast wave is **critical**). Intra-alveolar haemorrhage causes acute respiratory distress syndrome (*OHCM* p 176). Most patients who survive lung blast injury regain good lung function within a year.

n = 11
medline:10593795

2 Blast wind Air displaced by the explosion will totally disrupt a body in the immediate vicinity. Others may suffer avulsive amputations. Bodies can be carried by the wind with deceleration injuries on landing. Glass, wood, stones, and other objects are also carried and act as secondary missiles.

medline:10892035

3 Missiles Penetration or laceration from missiles are by far the commonest injuries. Missiles arise from the bomb (casing or preformed fragment—nails, nuts, and bolts), or are secondary (as above, glass and wood particularly).

4 Flash burns These are usually superficial and occur on exposed skin (hands/face) in those close to explosion.

5 Crush Injuries result from falling masonry.

6 Psychological Acute fear and panic is the aim of the terrorist. Later, chronic intrusive thoughts, anxiety, and poor concentration may form the basis of a post-traumatic stress disorder (p 347). This may be augmented by repeated watchings of the event on tv/video. medline:10686250

Treatment Approach the same as any major trauma with priority to airway and cervical spine control, breathing, and circulation with haemorrhage control. Rest and observe any suspected of exposure to significant blast, but without other injury. Sudden death or renal failure may follow release of a limb after prolonged crush (hyperkalaemia and myoglobinuria): ensure continuous ECG and adequate hydration. Facial burns may compromise airway, which should be secured by intubation or surgical airway (*OHCM* p 715). Psychological support will be required.

Diving accidents

Thousands go diving every year for recreation. Because of speedy world travel, complications of diving may present to doctors miles from diving centres: for this reason we all need to be familiar with the contraindications and complications of diving, *whatever* our specialty. Do not underestimate the stress of diving: a depth change of 7m produces changes in ambient pressure equal to a trip from sea level to the top of Everest.*

Contraindications to diving

- Migraine + vomiting attacks
- Otitis (media or externa)
- Hypoglycaemia risk (eg DM)
- Patent foramen ovale*
- Lung diseases
- Epilepsy
- Ménière's disease
- Pregnancy
- Pneumothorax history
- Angina; arrhythmias
- Perforated ear drum
- Bleeding disorders

Complications of diving

- Drowning (p 682)
- Marine bites or stings (p 736)
- Barotrauma
- Air embolism
- Hypothermia
- Decompression sickness
- Pulmonary oedema
- Nitrogen narcosis

Decompression sickness Nitrogen is more soluble in lipid than in water, so as N_2 tension increases, it accumulates in CNS, marrow and fat. Symptoms appear from 1–36h after surfacing. Risk factors: multiple dives, ignoring proper decompression stops, rapid ascent, previous decompression sickness, alcohol ingestion, subsequent ascent to high altitude (p 798).

Presentation:

- Vomiting
- Throbbing muscle/joint pains
- Migrating skin mottling/rashes
- Pruritus; paraesthesiae
- Mood changes
- Cough; chest pain
- Cyanosis; shock
- Osteonecrosis
- Deafness/nystagmus
- Fits; CNS signs
- Cognitive changes
- Headache

Management: Speed of response is vital. If recompression starts ≤ 30 min after the onset of symptoms, 80% will respond; if 6h delay, only 50% respond. If air embolism is suspected, place on the left side and give 100% O_2 (15L/min through tight-fitting face mask with reservoir). Seek expert help. Transfer to a hyperbaric facility.* If airlift needed, maximally pressurize the cabin, if the route does not allow flying at sea-level. Transport the breathing apparatus and his diving partner (will give the history, but may himself also be at risk). If there is hypothermia, expect BP to drop after leaving the water, and be sure to transport in the horizontal position, and keep cardiovascular instability to a minimum.

Preventing diving accidents

- Augment swimming fitness.
- Avoid dehydration (no alcohol or caffeine for >24h before the dive).
- Do the deepest part of the dive first. Time your ascent.
- Make a safety stop at 4.5m. Avoid remaining under water for longer than is recommended by decompression tables or dive computers.
- Rest before the dive, and keep warm during the dive.
- After the dive avoid sitting still for long periods (decreases regional blood flow and nitrogen removal). Avoid boarding aircraft for 12h.
- No diving if pregnant: there is (uncertain) risk to the fetus as right-to-left shunting diverts blood from the lungs—which are the best filters of micro-bubbles. (The rôle of the placenta in this task is unknown.) medline:10416137

*See M Knauth 1997 *BMJ* i 689 & 701 [09116544]: ► This careful MRI study in 160 asymptomatic scuba divers found many CNS lesions (?akin to multi-infarct dementia) associated with a patent foramen ovale (or other shunt) allowing paradoxical embolism of venous nitrogen bubbles (venous bubbles occur after ascent from as little as 3m (these bubbles are normally filtered by the lungs). ** Telephone numbers: UK: Hyperbaric Medicine Unit's diving doctor 01224 681818; Royal Navy: 01705 822351 or 01705 818888. USA: Diving Accident Network (919 684 8111) or us Navy 904 234 4351 or Air Force (512 538 3281 or 512 536 3278)

▶▶ Thrombolysis at home?

Time is muscle—once myocardial infarction is suspected, the chief goal is to relieve pain, correct arrhythmias, and to get patients to where thrombolysis can be started *urgently*. Only fire, exsanguination, or cardiorespiratory arrest should distract from ruthlessly pursuing this goal. We need to take steps to measure and reduce the time taken from contact with the patient to starting thrombolysis—the ‘door to needle’ time. This is not controversial: but the question now being asked is whose needle (GP or hospital?) and which door (the patient’s front door or the hospital door?). Should we assume that home use of thrombolysis would not much affect the outcome? Would such a strategy lead inevitably to the dangerous, inappropriate use of thrombolysis, and could it cause fatal delay in getting sick patients to hospital? The answer to all these questions is probably ‘No’ ($P < .04$)—in the light of a good randomized trial on UK patients. This shows that early thrombolysis (30u anistreplase i.v. over 5min, *OHCM* p 647) can halve mortality—and significantly improve all measures of outcome—compared with giving thrombolysis as fast as possible, once in hospital. medline:11428853 &: 1393033

In this trial, patients were given a twin pack of 2 injections, one was marked to be given at home and the other in hospital. One contained anistreplase and the other a placebo, and they were randomized between being the home injection or the hospital injection, so patients received two injections, one of which was active, and what was varied was its timing. The median delay between patient-contact and needle was 53min for general practitioners; patients had to wait a further 2h for the hospital injection, with most of this delay being within the hospital rather than in transit (despite the fact that the practices from which patients came were fairly remote from hospital (mean=36 miles). Almost no patient (0.6%) had a hospital injection within 2h of symptom onset, but 61% had their home injection within this time—and the subgroup having thrombolysis within this time did best of all (in other words, instead of a dose-response curve, there is a time-response curve).

Note that the GPs were as adept at diagnosing myocardial infarction as their hospital counterparts—the main difference being that their assessment was much faster (door-to-needle time of 43min compared with a calculated 87min). Note that only one patient admitted with a suspected coronary could be assigned with confidence to another diagnosis during the admission procedure. NB: criticisms of this study included the following: some patients may have had unstable angina, not infarction; medline:1458114 many possible patients may not have been recruited to the trial; and the number of deaths was small; making it possible that the lowering of mortality was exaggerated. medline:1458113 &: 1458116 &: 8326971 &: 1442633

▶▶ The vital point is that thrombolysis needs to be delivered with military precision and determination—with one part of our mind devoted to time-auditing our actions, and eliminating all non-essentials. Hence our uncharacteristic use of the word ‘ruthless’ above. Whether in general practice or in hospital, we have a duty to adapt our working practices to fast-track patients with chest pain to ensure they receive the care they need quickly. Do not think of this as a chore: it can be a luxury to free our minds and actions of the myriad distractions which ensnare an average day on call, and so save life.

Other care must also be prompt (*OHCM* p 104): reassurance, pain relief, arrhythmia correction, trinitrate spray, O₂, and the chewing of aspirin.

The last page

It is a pleasure to end this work with a chapter which is really a new beginning: the patient on his way to hospital. So far we have concentrated on what we can bring to the patient; but now let us turn to what the patient brings to us. All too often time and circumstance lead us to the view that patients are tireless devourers of our energies, and that for all practical purposes, we must go on giving until we die, or give up the unequal struggle with Nature and her diseases. This is to ignore the view that patients can be a source of nutrition: not just in the sense of providing us with our daily bread and butter, but also in the sense of nourishing our personalities. They do this by telling us about ourselves. You may think that you are kind and wise, or clumsy and inadequate, and it takes our patients to disabuse us of these illusions, and to show us that some days we are good, and some days we are bad. Thanks to our patients, we never stay the same. After practising medicine for a decade or two, our minds become populated by the ghosts of former patients, beckoning us, warning us, reminding us of the things we cannot control—and the ideals to which we aspire. We are lucky to work in a profession in which experience counts for more than knowledge, and it is to augment this thirst for experience that we urge our readers to turn away from learning by rote: let us read novels, cultivate our friends, travel far and wide—and try to keep forever curious, for then, if we are lucky, we stand to gain that priceless therapeutic asset: a rich and compassionate personality, and we will be all the more inclined to reformulate this tiresome and inconvenient patient who now confronts us into a lovable series of imperfections, which match and reflect, and reveal our own characteristics.

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Reference intervals—clinical chemistry

See p 292 for *children*; p 81 for *obstetric reference intervals*

►All laboratory discourse is probabilistic. ►Drugs may interfere with any chemical method; as these effects may be method-dependent, it is difficult for us to be aware of all possibilities. If in doubt, discuss with the lab.

Substance	Specimen	Normal value	Your hospital
Adrenocorticotrophic hormone	P	<80ng/L	
Alanine aminotransferase (ALT)	P	5–35iu/L	
Albumin	P††	35–50g/L	
Aldosterone	P**	100–500pmol/L	
Alkaline phosphatase	P††	30–300iu/L (adults)	
α-fetoprotein	S	<10ku/L	
α-amylase	P	0–180 Somogyi u/dL	
Angiotensin II	P**	5–35pmol/L	
Antidiuretic hormone (ADH)	P	0.9–4.6pmol/L	
Aspartate transaminase	P	5–35iu/L	
Bicarbonate	P††	24–30mmol/L	
Bilirubin	P	3–17μmol/L (0.25–1.5mg/100 /mL)	
Calcitonin	P	<0.1μg/L	
Calcium (ionized)	P	1.0–1.25mmol/L	
Calcium (total)	P††	2.12–2.65mmol/L	
Chloride	P	95–105mmol/L	
*Cholesterol	P	3.9–to <6mmol/L	
VLDL	P	0.128–0.645mmol/L	
LDL	P	1.55–4.4mmol/L	
HDL	P	0.9–1.93mmol/L	
Cortisol	P	a.m. 450–700nmol/L midnight 80–280nmol/L	
Creatine kinase (CK)	P	♂ 25–195iu/L; ♀ 25–170	
Creatinine (related to lean body mass)	P††	70–≤150μmol/L	
Ferritin	P	12–200μg/L	
Folate	S	2.1μg/L	
Follicle-stimulating hormone (FSH)	P/S	2–8u/L (luteal): ovulatory peak 8–15 follicular phase, & ♂: 0.5–5 postmenopausal: >30 ♂ 11–51; ♀ 7–33iu/L	
Gamma-glutamyl transpeptidase	P	♂ 11–51; ♀ 7–33iu/L	
Glucose (fasting)	P	3.5–5.5mmol/L	
Glycated (glycosylated) haemoglobin	B	5–8%	
Growth hormone	P	<20mU/L	
Iron	S	♂ 14–31μmol/L; ♀ 11–30	
Lactate dehydrogenase (LDH)	P	70–250iu/L	
Lead	B	<1.8mmol/L	
Luteinizing hormone (LH)	P/S	premenopausal: 3–13u/L follicular: 3–12 ovulatory peak: 20–80 luteal: 3–16 postmenopausal: >30	
Magnesium	P	0.75–1.05mmol/L	
Osmolality	P	278–305mosmol/kg	
Parathyroid hormone (PTH)	P	<0.8–8.5pmol/L	
Phosphate (inorganic)	P	0.8–1.45mmol/L	
Potassium	P	3.5–5.0mmol/L	
Prolactin	P	♂ <450 u/L; ♀ <600 U/L	
Prostate specific antigen	P	0–4 nanograms/mL	
Protein (total)	P	60–80 g/L	
Red cell folate	B	0.36–1.44 μmol/L (160–640 μg/L)	

Renin (erect/recumbent)	P**	2.8–4.5/1.1–2.7 pmol/mL/h
Sodium	P†	135–145 mmol/L
Thyroid-binding globulin (TBG)	P	7–17 mg/L
Thyroid-stimulating hormone (TSH) NR widens with age,	P	0.5–5.7 mU/L
Thyroxine (T ₄)	P	70–140 nmol/L
Thyroxine (free)	P	9–22 pmol/L
Total iron binding capacity	S	54–75 µmol/L
Triglyceride	P	0.55–1.90 mmol/L
Tri-iodothyronine (T ₃)	P	1.2–3.0 nmol/L
Urea	P†	2.5–6.7 mmol/L
Urate	P†	♂ 210–480 µmol/L ♀ 150–390 µmol/L
Vitamin B ₁₂	S	0.13–0.68 nmol/L (>150 ng/L)

ARTERIAL BLOOD GASES

pH: 7.35–7.45	P _a CO ₂ : 4.7–6.0 kPa
P _a O ₂ : >10.6 kPa	Base excess: ± 2 mmol/l
NB: 7.6 mmHg = 1 kPa (atmospheric pressure ≈ 100 kPa)	

**The sample requires special handling: contact the laboratory

† Range is significantly different in pregnancy, see p81

Key: P=plasma (heparin bottle); S=serum (clotted; no anticoagulant); B=whole blood (edetic acid—EDTA—bottle); IU=international unit ♂ = male; ♀ = female.

Haematology—reference intervals

Measurement	Reference interval	Your hospital
White cell count (wcc)	4.0–11.0 × 10 ⁹ /L	
Red cell count	♂ 4.5–6.5 × 10 ¹² /L ♀ 3.9–5.6 × 10 ¹² /L	
Haemoglobin	♂ 13.5–18.0 g/dl ♀ 11.5–16.0 g/dl	
Packed red cell volume (PCV) or haematocrit	♂ 0.4–0.54 l/l ♀ 0.37–0.47 l/l	
Mean cell volume (MCV)	76–96 fl	
Mean cell haemoglobin (MCH)	27–32 pg	
Mean cell haemoglobin concentration (MCHC)	30–36 g/dl	
Neutrophils	2.0–7.5 × 10 ⁹ /L; 40–75% wcc	
Lymphocytes	1.3–3.5 × 10 ⁹ /L; 20–45% wcc	
Eosinophils	0.04–0.44 × 10 ⁹ /L; 1–6% wcc	
Basophils	0.0–0.10 × 10 ⁹ /L; 0–1% wcc	
Monocytes	0.2–0.8 × 10 ⁹ /L; 2–10% wcc	
Platelet count	150–400 × 10 ⁹ /L	
Reticulocyte count	0.8–2.0%* 25–100 × 10 ⁹ /L	
Erythrocyte sedimentation rate	depends on age (OHCM p 672)	
D-dimer	<0.5 mg/L (check with lab)	
Activated partial thromboplastin time (VIII, IX, XI, XII)	35–45 seconds	
Prothrombin time (I, II, VII, X)	10–14 seconds	

Internat. ratio (INR)	Clinical state (see OHCM p 650–1)
2.0–3.0	Treating DVT, pulmonary emboli (treat for 3–6 months)
2.5–3.5	Embolism prophylaxis in atrial fibrillation, see OHCM p 651
3.0–4.5	Recurrent deep vein thrombosis and pulmonary embolism; arterial disease including myocardial infarction; arterial grafts; cardiac prosthetic valves (if caged ball aim for 4–4.9) and grafts.

Reference intervals—biochemistry

See p 292 for *children* and p 81 for *obstetric reference intervals*

►All laboratory discourse is probabilistic. ►Drugs may interfere with any chemical method; as these effects may be method-dependent, it is difficult for us to be aware of all possibilities. If in doubt, discuss with the lab.

Substance	Specimen	Normal value	Your hospital
Adrenocorticotrophic hormone	P	<80ng/L	
Alanine aminotransferase (ALT)	P	5–35iu/L	
Albumin	P††	35–50g/L	
Aldosterone	P**	100–500pmol/L	
Alkaline phosphatase	P††	30–300iu/L (adults)	
α-fetoprotein	S	<10ku/L	
α-amylase	P	0–180 Somogyi u/dL	
Angiotensin II	P**	5–35pmol/L	
Antidiuretic hormone (ADH)	P	0.9–4.6pmol/L	
Aspartate transaminase	P	5–35iu/L	
Bicarbonate	P††	24–30mmol/L	
Bilirubin	P	3–17μmol/L (0.25–1.5mg/100 /mL)	
Calcitonin	P	<0.1μg/L	
Calcium (ionized)	P	1.0–1.25mmol/L	
Calcium (total)	P††	2.12–2.65mmol/L	
Chloride	P	95–105mmol/L	
*Cholesterol (see p654)	P	3.9–7.8mmol/L	
VLDL (see p654)	P	0.128–0.645mmol/L	
LDL	P	1.55–4.4mmol/L	
HDL	P	0.9–1.93mmol/L	
Cortisol	P	a.m. 450–700nmol/L midnight 80–280nmol/L	
Creatine kinase (CK)	P	♂ 25–195iu/L; ♀ 25–170	
Creatinine (related to lean body mass)	P††	70–≤150μmol/L	
Ferritin	P	12–200μg/L	
Folate	S	2.1μg/L	
Follicle-stimulating hormone (FSH)	P/S	2–8u/L (luteal): ovulatory peak 8–15 follicular phase, & ♂: 0.5–5 postmenopausal: >30	
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P _a O ₂ : >10.6 kPa	Base excess: ± 2 mmol/l
	NB: 7.6mmHg=1kPa (atmospheric pressure≈100kPa)

- * Desired upper limit of cholesterol would be ~6mmol/l.
- **The sample requires special handling: contact the laboratory
- †† Range is significantly different in pregnancy, see p81

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Prothrombin time (factors I, II, VII, X)	10–14 seconds	

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2.5–3.5	Embolism prophylaxis in atrial fibrillation, see OHCM p 597
3.0–4.5	Recurrent deep vein thrombosis and pulmonary embolism; arterial disease including myocardial infarction; arterial grafts; cardiac prosthetic valves (if caged ball aim for 4–4.9) and grafts.

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